	BCG scar is present **Ear, Nose, throat** examination is unremarkable		**loud S2. JVP is not raised.** **No signs of steroid toxicity**
Differentials	CONSOLIDATION 1) **Pneumonia** 2) **Tuberculosis** 3) **Collapse with patent main bronchus**	PLEURAL EFFUSION 1) **Parapneumonic** 2) **Post Tuberculosis** 3) **Empyema** 4) **Collapse with obstructed main bronchus**	BRONCHIECTASIS 1) **Post tuberculosis** 2) **Post pneumonic** 3) **Post viral** (Pertussis, measles) 4) **Cystic fibrosis** 5) **Primary ciliary dyskinesia** 6) **Immunodeficiency** 7) **Congenital lung lesion** 8) **Interstitial lung disease**
Investigations	1) **CBC** for leukocytosis 2) **CXR** for consolidation/effusion 3) **Mantoux test** for TB 4) **Gene Xpert** FOR TB 5) **Sputum culture** (limited value) 6) **Blood culture** (if severe disease) 7) **Nasopharyngeal aspirate** (viral immunofluorescence in infants) 8) **Pleural fluid** (culture and antigen testing)		1) **CBC** for leukocytosis 2) **CXR** (nonspecific) 3) **Mantoux test** for TB 4) **Gene Xpert** FOR TB 5) **Sputum culture** (limited value) 6) **Sweat test** 7) **Immunologic workup** 8) **Pulmonary function studies** 9) **Thin-section HRCT scanning**
Treatment	1)**MDT** 2)**Parental counselling** 3)**Supportive care** 4)**Antibiotic therapy** : (for 7-10 days , 14-21 days if Staphylococcus) **Under 5yrs**: Amoxicillin, Co-amoxiclav, Cefaclor **Over 5yrs**: Erythromycin, Clarithromycin, Azithromycin **For Staph Aureus:** Flucloxacillin with amoxicillin **Severe pneumonia** : IV Co-amoxiclav/cefotaxime/ cefuroxime **Nosocomial Pneumonia** : 3 Drugs Vancomycin Piperacillin-tazobactum / Meropenem Gentamicin	1)**MDT** 2)**Parental counselling** 3)**Supportive care** 4)**Antibiotic therapy** IV Anti biotics for 2-4 weeks 5)**Surgical treatment** a) Chest intubation for 1 week b) VATS followed by chest tube c) Instillation of fibrinolytic agent: (Streptokinase/Urokinase) d) Thoracotomy (VATS/Open surgery)	1)**MDT** 2)**Parental counselling** 3)**Supportive care** 4) **Medical therapy** a) IV Antibiotics (2- 4 weeks in acute exacerbations) b) Bronchodilators c) Chest physiotherapy d) Corticosteroids (inhaled) e) Treat the cause (immunodeficiency, aspiration) 5)**Surgical treatment** a) Segmental or lobar resection b) Lung transplantation

NOTES

RESPIRATORY DISEASES
(DESCRIPTION & VIVA)

Opening	Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in place.		
	CONSOLIDATION	EFFUSION	BRONCHIECTASIS
General look	He is **pink in room air** with no **signs of distress** (with signs of respiratory distress in the form of subcostal recessions and nasal flaring) or **dysmorphism.**		He is **peripherally cyanosed, clubbed ± Signs of respiratory distress** (tachypnea, recessions and nasal flaring)
Vitals	He is afebrile (to touch), Respiration is abdominothoracic with Respiratory Rate ___/min, Pulse is ___/min Regular in rhythm and normal in Volume & Character; B.P. is _____mmHg.		
Inspection	Chest is **normal in shape** with no scars, bulging, prominent veins or Harrison sulcus	Chest is **normal in shape/Left/Right hemithorax is bulging** with no scars, , prominent veins or Harrison sulcus	Chest is **Barrel shaped** with no scars, prominent veins or Harrison sulcus.
Palpation	Chest is **non tender** & there is No evidence of **mediastinal shifting & dextrocardia**	Chest is **non tender** & there is No evidence of **mediastinal shifting & dextrocardia** *(Effusion can push mediastinum)*	Chest is **non tender** & there is No evidence of **mediastinal shifting & dextrocardia**
My findings are confined to _____(area) in the form of			
Expansion	**Decreased chest expansion**	**Decreased chest expansion**	**Decreased chest expansion**
Vocal Fremitus	**Increased vocal fremitus**	**Decreased vocal fremitus**	**Normal/Increased vocal fremitus**
Percussion	**Dull percussion note**	**Stony dull percussion note**	**Normal/Dull percussion note**
Auscultation	**Bronchial breathing** with crackles	**Decreased Breath sounds** ,no added sounds	**Vesicular/ Bronchial breathing with bilateral crackles/ronchi**
Vocal Resonance	**Increased vocal resonance**	**Decreased vocal resonance**	**Normal/Increased vocal resonance**
Abdomen	There is no **hepatosplenomegaly & Pedal edema**		
Back	Back is **Non tender & normal in shape with no visible deformity. Similar findings** are found in _____(area)		
GPE	There is no evidence of **cyanosis, clubbing,**pallor & **lymphadenopathy**	All previous + **No oral ulcer,rash**	All previous + **Both heart sounds are audible with**

	Teeth: Age appropriate, dental caries, abnormal teeth **Tonsils**: inflamed/pustular exudates on tonsils (tonsillitis) **Pharynx**: Red and inflamed (pharyngitis) **Lymph nodes:** Occipital, postauricular (from front)
Abdomen	**Liver**
Lower limb	**Toenail clubbing** **Ankle oedema** (right-ventricular failure)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) **B.P.**
2) **Fundoscopy** : Retinal venous dilatation (CO_2 retention)
3) **Otoscopy**
4) **Anthrpometric measurements**
5) **Neurological system** for predisposing causes of respiratory distress, e.g. bulbar palsy
6) (Aspiration), spinal muscular atrophy (diaphragmatic breathing)

TIPS & TRICKS

1) Don't forget apices.
2) Keep stethoscope on an ICS for one complete respiratory cycle.
3) Cyanosis is easily missed if pink curtains surround child's bed.
4) Describe findings in reference to specific surface locations.
5) Adjust your findings according to your first differential e.g. if your differential is consolidation do not say vocal resonance is normal or decreased.
6) Listen to the command and proceed accordingly

'Examine the respiratory system'	'Examine the chest'
Includes the hands (starting with the nails for clubbing), chest wall, precordium, lungs, ears, nose, throat and regional lymph nodes.	Chest wall (starting with this, not the hands), heart and lungs, as the primary focus

NOTES

3) Locate Apex beat (initially with both hands to rule out dextrocardia/displacement)
4) Chest expansion (at two places from front)
5) Vocal fremitus (Can be skipped if time is less)

STEP IV: PERCUSSION

Percuss upper border of liver (Can be Skipped in short case if time is less)
Percuss Intercostal spaces

Long Case : **648**	Short case : **325**
6 Spaces from front	**3** Spaces from front
4 from side	**2** from side
8 from back	**5** from back

STEP V: AUSCULTATION

Chest (in areas described above)
Heart (Time with carotid) (Look for Loud P2)
Vocal resonance *('333 bolain')*

STEP VI: BACK (Make child sit; back facing towards you and his arms crossed; hands placed on opposite shoulder)

1) **Inspection**
2) **Palpation** (Chest expansion, sacral edema, deformity, Tenderness)
3) **Percussion**
4) **Auscultation**

STEP VII: RELEVANT GPE (Make him sit)

Nails	**Clubbing** (CF, suppurative lung disease, pulmonary fibrosis, coexistent cyanotic heart disease), **Peripheral cyanosis**
Fingertips	Prick marks (BSL testing in CF patients with diabetes)
Palm	**Crease pallor** (anemia), **single palmar crease** (Trisomy 21)
Hands	**Dorsum of hand:** scars of previous multiple IVs **Joints:** swollen (HPOA) **Pulse :** Tachycardia (hypoxia, fever, treatment with beta-2 agonists), Pulsus paradoxus (e.g. in severe asthma) **Asterixis :** CO_2 retention
Arm	**BCG Scar, Axillary lymph nodes**
Head & Face	**Head** Size : Microcephaly/macrocephaly Shape: Dolicocephaly (ex-preterm=BPD) **Face:** Dysmorphic, Cushingoid (treatment with corticosteroids) **Eyes:** Conjuctiva (Palpebral/bulbar:Pallor)
ENT	**Ears**: otitis media, acute or chronic serous **Nose**: polyps (CF), DNS, Swollen turbinates **Sinuses**: Palpate for sinusitis? (But ask for tenderness in advance) **Mouth**: Halitosis (bronchiectasis), Cleft lip, palate **Tongue**: color: Pallor, cyanosis, jaundice, mucous membranes: ulcers, candidiasis

RESPIRATORY SYSTEM EXAMINATION

1) General Look
2) Inspection of Chest
3) Palpation
4) Percussion
5) Auscultation
6) Back
7) Relevant GPE

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building** (Talk to child for hoarseness)
c. **Position patient** [**45 degree supine & arms abducted**]
d. **Exposure:** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Most important thing: Look for clubbing

Clubbing: Likely Bronchiectasis	**No Clubbing**: Likely pleural effusion/consolidation

Sick/healthy looking
Obese/Thin / malnourished
Dysmorphic features
Eczema (e.g. with asthma)

STEP II: INSPECTION OF CHEST

From Foot end	**Respiratory Rate** (Count for 10 sec x 6) (Abdominothoracic respiration) **Respiratory Noises** (stridor or wheeze) **& Coughing** **Rhythm** : Regular/periodic(infants) **Pattern**: Abdominothoracic (kids/male adults) (Thoracic breathing=Intra-abdominal pathology) **Signs of Respiratory distress** **Chest deformity, Asymmetry, Scars, Venous access devices**
From Right side	**Reinspect** from side & from above shoulder (for anything missed)(barrel chest, Harrison's sulcus, pectus carinatum or excavatum) **Ask the child to cough**

STEP III: PALPATION

Warm your hands and Ask: *'Chaatti main kahiin daard tuu nahi?'*
1) Superficial palpation (For subcutaneous emphysema/Tenderness)
'Maiin apki saans ki naali mehssos karoon ga iss say apko dard nahi ho ga!'
2) Palpate trachea (three finger method)

TIPS & TRICKS

1) Command can be GPE or Abdomen so start accordingly with main focus on command.

NOTES

2) Evaluation of kidney function
3) Quantitation of urine protein excretion

Causes : FSGS (80%), MCNS, or membranoproliferative glomerulonephritis

FSGS is associated with a 50% risk for ESRD within 5 yr of diagnosis if patients do not achieve a partial or complete remission.

Mendoza protocol: IV pulse methylprednisolone 600mg/m^2 for 82 weeks

Alternate day	2 weeks	
Weekly	8 weeks	Start oral predinsolone 2mg/kg on alternate day
Alternate week	8 weeks	
Monthly	8 months	
2 Monthly	8 months	

13) ESRD needing Dialysis / kidney transplantation (esp FSGS pts)

Steroid-resistant FSGS ➔ Around 8–10% develop ESRD

Recurrent nephrotic syndrome develops in 30-50% of transplant recipients with FSGS.

Complications of treatment

Cushingoid effects (Steroids)

Poor growth ** (Most serious complication) (Steroids)

V. MONITORING OF DISEASE & THERAPY

1) **Home urine protein**
2) **Anthropometry, GPE, BP**
3) **RFTs & Electrolytes**
4) **LFTs** (Mycophenolate, HMG Co-A inhibitors)
5) **CBC**
6) **BSR**
7) **Lipid profile**
8) **CPK** (HMG Co-A inhibitors)
9) **Drug levels** (Calcineurin inhibitors)

PROGNOSIS & COUNSELLING

Brief about pathophysiology, treatment , signs and symptoms of the complications & treatment

1) How to use and interpret the results of urinary dipstick testing for protein
2) Seek medical attention if the child appears ill, has a fever, or complains of persistent abdominal pain.

The best prognostic indicator in NS is steroid sensitivity. (Must tell)

1) Steroid-responsive NS is unlikely to develop chronic kidney disease
2) Steroid-responsive NS is Rarely hereditary
3) Child (in the absence of prolonged cyclophosphamide therapy) will remain fertile.
4) Children with idiopathic nephrotic syndrome should not be considered chronically ill and should participate in all age-appropriate childhood activities.

CSINS	CRINS
60–80% will relapse (few relapses if >4yr & no microhaematuria)	Steroid-resistant FSGS, many progress to ESRD = Renal transplantation
60% of them have five or more relapses	FSGS recurs = 25% of renal allografts
At 10 yr non-relapse rate = 84%	

(Avoid drinking grapefruit juice with statins= increase blood levels of statins)

5) Hypertension

ACE inhibitors or ARBs (as these delay deterioration in renal function)

6) Obesity

Anticipatory dietary counseling
Steroid-sparing strategies

7) Negative nitrogen balance

Loss of protein in urine, plus poor appetite, and nausea contribute to poor intake.

8) Growth failure (esp in congenital nephrotic syndrome)

Cause: Urinary loss of insulin-like growth factor binding protein (IGFBP) ➔ decreased IGF-I , IGF-II, and IGF-receptor mRNA
Use of corticosteroids
Rx
Steroid-sparing strategies
Recombinant human GH (rhGH) (?)

9) Hypothyroidism (esp in congenital nephrotic syndrome)

Cause : Loss of thyroid binding protein in the urine

10) Hypocalcaemia

Cause: Loss of vitamin D-binding protein in the urine ➔ Bone demineralisation in the long term

11) Relapse/Steroid dependence

60 mg/m^2/day (daily/TDS) until patient is free of proteinuria for 3 days
40mg/m^2/day OD on alternate days for 6 weeks
Gradually taper every 2 weeks by 15mg/m^2

Other agent

Levamisole (Tab Ketress 40mg = 7rs, Syp 40mg/5ml :15ml=34rs) (@ 2.5mg/kg on alternate day) for 12–24 months *(Aj's crack : 15kg = 1 tablet/1 tsf on alternate day)* + **low dose steroids** (Pre-school: 1 mg/kg/alternate day, School going: 0.5 mg/kg/alternate day)
Mycophenolate mofetil (Tab Cellcept 500mg : 160 Rs)(25 mg/kg/day in divided doses for 12–24 months) *(Aj's crack : 10 kg = 1/4 tablet x BD)*
Cyclophasphamide (Endoxan Tab 50mg = 7rs, Inj 500mg= 151rs) + alternate day steroids ***Oral*** : 2mg/kg daily orally 8-12 weeks morning dose ***IV*** : 500mg/m^2 monthly for 6 months
Cyclosporin A (Sandimmune Inj 250mg/5ml=520 Rs, 100mg/ml sol 50ml=6951rs) (2.5mg/kg/day) for 12-24 months + low dose steroids

12) Steroid resistance (No remission in 8 weeks)

Further evaluation

1) Diagnostic kidney biopsy

(Give only if prednisone dose is below either 1 mg/kg daily or 2 mg/kg on alternate days)
Vaccines can be administered after corticosteroid therapy has been discontinued for at least 1 month. (Pneumococcal, Haemophilus and varicella vaccines are recommended)
Influenza vaccine should be given on a yearly basis.(to the child and their household contacts)
Immunize healthy household contacts with live vaccines to minimize the risk of transfer of infection to the immunosuppressed child, but avoid direct exposure of the child to GI or respiratory secretions of vaccinated contacts for 3-6 wk after vaccination.
Exposure to Varicella in Non-immune nephrotic children in relapse : Give varicella-zoster immunoglobulin (1 dose ≤96 hours after significant exposure)

3) Thromboembolic events (2-5%,lower risk than in adults)

Most commonly : Renal vein thrombosis OR sagittal sinuses thrombosis
Others: deep vessels of the limbs, the pulmonary artery, the inferior vena cava, the femoral/iliac artery, pulmonary venous system, the cerebral arteries, the meningeal arteries, mesenteric veins and the hepatic veins, thrombosis of indwelling arterial and venous catheters

Thromboembolic complications
Pulmonary embolism
Bilateral renal vein thrombosis ➔ AKI (Esp. in congenital nephrotic syndrome)

Risk factors
CRINS
Congenital nephrotic syndrome
Coexisting illness leading to fluid loss
Haemoconcentration (Vomiting or diarrhea)
Diuretic use
Immobilisation
Presence of indwelling catheters

Rx
Avoid aggressive use of diuretics and the use of indwelling catheters
First line : LMWH (if a previous thromboembolic event)
If the thrombosis extends: Thrombolytic drugs (e.g. tissue plasminogen activator) + Warfarin until the nephrotic syndrome resolves
Aspirin therapy (can prevent arterial thrombosis Not venous:routine use not recommended)

4) Hyperlipidemia/Dyslipidemias & cardiovascular disease risk

Hyperlipidaemia: reversed quite quickly in CSINS (4–6 weeks)

Risk factors for cardiovascular sequelae
Exposure to corticosteroids
Hypertension
Hypercoagulability
Anaemia (erythropoietinresponsive anaemia)

Rx
Low-fat diet (<30% of calories with saturated fat intake <10% calories)
Dietary cholesterol intake should be <300 mg/day.
3-hydroxy- 3-methylglutaryl coenzyme A (HMG-CoA) reductase-inhibiting drugs
Tab Lovastatin (Exelip,Clolip 20mg) FDA approved* ½ OD
Tab Atorvastatin(Lipiget 10mg) 1 x OD

ACE inhibitors	
Tacrolimus	
Methyl Pred?	IV methylprednisolone @ 30 mg/kg, Thrice a week (for 2 weeks) then tapering gradually over 80 weeks + concurrent prednisolone on alternate days (Advantage: remission rates of 60–70%) (Very toxic: hypertension, delayed growth, cataracts, infections)
Mycophenolate ?	
Rituximab ?	Chimeric anti-CD20 monoclonal antibody

Patients Not remitting usually progress to ESRD. Thus it is important to try therapies to achieve complete or partial remission, but without excessive medication-related toxicity.

IV. TREATMENT OF COMPLICATIONS

A) Disease related	B) Related to treatment
1) Edema 2) Infection 3) Thromboembolic events	Cushingoid effects (Steroids) Poor growth (Steroids) Cataract
4) Hyperlipidemia & cardiovascular disease risk 5) Hypertension 6) Obesity 7) Negative nitrogen balance	BM suppression (Cyclophosphamide) Viral infections Hemorrhagic cystitis Gonadal toxicity & carcinogenesis
8) Growth failure 9) Hypothyroidism 10) Hypocalcaemia	Nephrotoxicity, Hypertension, Gingival hyperplasia & hypertrichosis (Cyclosporine A)
11) Relapse 12) Steroid resistance 13) ESRD needing Dialysis / transplantation	Gastrointestinal effects & Haematological abnormalities (Mycophenolate mofetil)

1) Edema

Anasarca (generalised, massive oedema) can lead to the following:
Difficulty walking (Severe scrotal or vulval oedema)
Respiratory distress (Pleural effusions and/or severe ascites splinting the diaphragm)
Tissue breakdown with cellulitis

2) Infection *** (SBP*** (2–6%), meningitis , sepsis, pneumonia, cellulitis, UTI)

Antibiotics

Organisms for SBP(Spontaneous bacterial peritonitis): Streptococcus pneumonia***, E.coli
A high index of suspicion for bacterial peritonitis (fever, abdominal pain, and peritoneal signs), prompt evaluation (Blood & peritoneal fluid cultures) and early antibiotic therapy are critical.
Peritoneal leukocyte counts >250 are highly suggestive of spontaneous bacterial peritonitis.

Prophylactic daily penicillin (To avoid pneumococcal infection during relapse)
3rd Generation cephalosporin (e.g. cefotaxime) (Penicillin-resistant pneumococcus/coliforms) + Ampicillin (to cover for Enterococcus)

Immunization

AVOID LIVE VACCINES (While on high-dose corticosteroids/immunosuppressive agents)

CORTICOSTEROID-SENSITIVE/DEPENDENT IDIOPATHIC NS (CSINS)

Sensitivity **CLAIMING** drugs : **CLAIM**

C	Cyclophosphamide (CPA: 2 mg/kg/day for 8–12 weeks) + alternate day steroids Chlorambucil (0.2 mg/ kg/day day for 8 weeks) Cyclosporine A (CSA: 2.5 mg/kg 12-hourly for 12–24 months)
L	**L**evamisole (2.5 mg/kg alternate daily for 12–24 months)
A	**A**ngiotensin-converting enzyme (ACE) inhibitors
I	**I**mmunisation with pneumococcal vaccine
M	**M**ycophenolate mofetil (25 mg/kg/day in divided doses for 12–24 months)
	Rituximab ? Chimeric anti

Indications

1) Failure to respond to steroids (Relapse on prednisolone dosage > 1mg/kg/day)
2) Unacceptable steroid side effects (e.g. reduced height gain)
3) Steroid-dependent NS (only if side effects are unacceptable)
4) Relapses with hypertension / thrombosis (if frequent relapse/steroid dependent)

Poor compliance is not an indication, as monthly IV methylprednisolone can be given

Levamisole (1st)	Helps maintain remission in steroid-dependent INS ↓ Steroid requirement 50% ↓ Relapse rate by 50%	Few side effects (reversible) Neutropenia Liver toxicity Convulsions, Rarely vasculitis
Mycophenolate mofetil (2nd)	Efficacy = Cyclosporine Less side effects than CSA	Gastrointestinal effects (diarrhoea and abdominal pain) & Haematological abnormalities
Cyclophosphamide CPA (3rd) Relative contraindication = Lack of Varicella antibodies	Prolonged remission off therapy (5 year remission for frequently relapsing CSINS = 36%) Sustained effect Given for 8-12 weeks(2–4 month) Monitor WBC count weekly (Stop if count < 5,000/mm^3)	Significant side effects **Short term** (e.g. BM suppression, Risk of viral infections e.g. Varicella, Measles) **Long term** (e.g. Hemorrhagic cystitis, alopecia , Gonadal toxicity & risk of carcinogenesis) *Cumulative threshold dose above which oligo- or azoospermia occurs in boys is >250 mg/kg*
Cyclosporine A CSA (4th)	Not sustained effect Given for 12–24 months	Nephrotoxicity, Hypertension, Gingival hyperplasia & hypertrichosis

*While a patient is taking these drugs, the full blood count must be checked regularly

Pt on cyclosporine, tacrolimus, or mycophenolate therapy tend to relapse when the medication is discontinued.

CORTICOSTEROID-RESISTANT IDIOPATHIC NEPHROTIC SYNDROME (CRINS)

CAT resists Corticosteroids = **CAT** drugs

Calcineurin inhibitors (cyclosporine or tacrolimus) are initial therapy for CRINS

Cyclosporine A (CSA)	Cyclosporine A **Alone** Cyclosporine A + IV Methylprednisolone for three doses

Temp. & others = Tepid sponging + Paracetamol
Tubes : NG, Foley's Catheter

Nutrition = At the initial presentation and during relapse, a low-sodium diet
No fluid restriction, except when very oedematous and diuretics are required.
High biological value protein.
Low-fat diet (<30% of calories with saturated fat intake <10% calories)
Dietary and fluid restrictions not required in remission.
Adequate **calcium and vitamin D intake.**
Healthy eating regimen + exercise (to prevent excessive weight gain)
Monitoring :

1) Fluid balance chart
2) Abdominal Girth
3) 12 hrly weight
4) 4-hourly temperature, pulse, respirations and blood pressure

Care of comatosed (EMA's BB Posture) : Eye, Mouth, Airway, Skin, Bowel, Bladder, Posture

III. SPECIFIC THERAPY

Age 1-8 yr & features of MCNS = Start treatment without Renal Biopsy
Age <1, >12 yr & features less likely of MCNS = Renal biopsy before treatment

Important Check before giving Steroids
Rule out T.B. before giving steroids (PPD, Chest X-ray, PPA Score)

1) Administer polyvalent pneumococcal vaccine

Conjugated pneumococcal vaccine (PCV 10,13) in children under 5	Polysaccharide pneumococcal vaccine (PPV23) in children 5 or older

2) Tab Prednisolone (Deltacortil) (5mg) (Syrup Steron 15mg/5ml)

60 mg/m²/day (maximum daily dose, 80 mg) X **OD** 2mg/kg/day	For 6 weeks (4-6weeks)	
40 mg/m²/day X OD on alternate day (1.5mg/kg/day)	For 4-6 weeks	Total 8 wk - 5 month
Taper alternate dose	Over 6 weeks	

3) Syrup Ranitidine (Peptinil 15mg/ml) 2-4mg/kg/day (10 kg = 2ml X OD)
OR Syrup Sucralfate (Ulsanic 100mg/ml) 40-80mg/kg/day q6hr (10kg = 2ml X TDS) (Before meal)

4) Angiotensin-converting enzyme (ACE) inhibitors and angiotensin II blockers may be helpful as adjunct therapy to reduce proteinuria in steroid-resistant patients
Tablet Captopril (Capoten 25mg) 1mg/kg/day divided BD (10kg = 1/5th BD)
A.E. : Hypotension and cough

Proteinuria (2+ or greater) after 8 wk of steroid therapy = Steroid resistant...Do diagnostic renal biopsy

One can follow KDIGO guidelines or following scheme for treatment of special cases.

SGN.....↓C3 persistent = Type 2 MPGN)
5) **HepBsAg** (associated with membranous nephritis)
6) **Anti HCV Ab** (associated with MesPGN)
7) **HIV serology** (if at risk)
8) **Torch screening** (congenital nephrotic)

III. TO RULE OUT COMPLICATIONS

1) **Urine C/S**
2) **Serum Urea Creatinine** (Normal in INS, abnormal in other GN)
3) **Serum Electrolytes** (Hyponatremia , ADH↑)
4) **Ca, Mg, PO4** (↓ Ca due to vit D binding protein are↓)
5) **TSH, T4**
6) **USG Abdomen** (Ascites)
7) **PPD** (Mantoux test)
8) **Chest X-ray** (Pleural effusion/TB)

TREATMENT

) Tabulated Overview

MDT	Supportive R_x	Specific R_x	R_x of Complications	
	ALP – OF - TTNM Care	Pneumococcal vaccine Steroids H2 receptor blockers ACEi Immunosuppressants	Edema Infection Thromboembolic events Hyperlipidemia HTN	Obesity Negative Nitrogen balance Growth failure Hypothyroidism Hypocalcemia

b) Details

I. MDT (Multi-disciplinary team approach)

Pediatrician	P. Nephrologist	Endocrinologist	Dietician
Physical therapist	Psychiatrist	Social worker	Nurse clinician

II. SUPPORTIVE TREATMENT (ALP – OF - TTNM Care)

Admit Children with severe symptomatic edema(large pleural effusions, ascites, or severe genital edema)

List
Position/ Placement :Prop up if respiratory distress, Elevate swollen scrotum with pillows
Oxygen:
Fluids & electrolytes : Sodium restriction (<1500 mg daily), fluid restriction (if hyponatremic)
Diuresis: Inj furosemide(Lasix), 1-2mg/kg/dose OR
Tablet Spironolactone+Furosemide(Spiromide 20mg) 1mg/kg/dose X BD (10kg= ½ Tab BD)
(Alert! Intravascular Fluid depletion, Risk of intravascular thrombosis)
If massive Edema+Intravascular volume depletion (↓B.P., ↑ H.R.) = Salt free Inj Albumin 20% (0.5-1g/kg/day)(5ml/kg) slow infusion over 4 hrs + Inj furosemide (1-2mg/kg/dose) after the first hour *(With monitoring of volume status, B.P., serum electrolyte balance, and renal function)*
(Complications of rapid Albumin infusion = volume overload, with hypertension, heart failure, and pulmonary edema)

DIFFERENTIALS

1) Nephrotic syndrome
2) Nephritic syndrome i.e AGN
3) To rule out CLD, CCF, Protein losing enteropathy

INVESTIGATIONS

a) Tabulated Overview

I. FOR DIAGNOSIS	II. ETIOLOGY	III. TO RULE OUT COMPLICATIONS
1) Urinalysis 2) Early morning U P:C ratio 3) Urinary protein excretion over 24hr 4) Serum albumin 5) Total protein levels 6) Lipid Profile 7) Renal biopsy 8) Genetic studies	1) CBC 2) MP slide 3) ANA ,double-stranded DNA 4) Serum complement levels 5) HepBsAg 6) Anti HCV Ab 7) HIV serology 8) Torch screening	1) Urine C/S 2) Serum Urea Creatinine (Normal in INS, abnormal in other GN) 3) Serum Electrolytes (Hyponatremia , ADH↑) 4) Ca, Mg, PO4 (↓ Ca due to vit D binding protein are↓) 5) TSH, T4 6) USG Abdomen 7) PPD (Mantoux test) 8) Chest X-ray

b) Details

I. FOR DIAGNOSIS

1) **Urinalysis**: 3+ or 4+ proteinuria , microscopic hematuria (20%), cellular casts for other glomerulonephropathies (hyaline or waxy casts are common in INS)
2) **Early morning Spot urine protein:creatinine ratio** > 2.0
3) **Urinary protein excretion over 24hr** > 40 mg/m^2/hr (not very useful)
4) **Serum albumin** level <2.5 g/dL (to evaluate severity)
5) **Total protein levels** (to evaluate severity)
6) **Lipid Profile** (Cholesterol and triglyceride levels are elevated) > 200

   Selected Cases
7) **Renal biopsy** if
   - Age <1 yr
   - Age >8 yr (esp. African Americans, where FSGS is more common)
   - Nephritic features (gross hematuria, hypertension)
   - Renal insufficiency/ raised creatinine
   - Persistent hypocomplementemia (low C3)
   - Steroid resistance
8) **Genetic studies**

II. TO FIND ETIOLOGY/SUPPORTIVE

1) **CBC** (Hb normal in INS/due to volume contraction↑ Hb,PLT,Hct… If anaemia other diagnoses to be considered)
2) **MP slide**
3) **ANA ,double-stranded DNA** (SLE) (not necessary in younger patients)
4) **Serum complement levels** (low with MesPGN, SLE; normal in INS…↓C3 transient Post

STEP III: ABDOMEN, CHEST & CVS

Abdomen	Inspect (transplant scar, caput medusa) Palpate • Hepatomegaly (CCF, CLD) • Splenomegaly (CLD) • Nephromegaly (polycystic disease, hydronephrosis) • Inguinal lymphadenopathy (if leg oedema only: local cause) Percuss bladder Demonstrate ascites (shifting dullness, fluid thrill)
Scrotum	Scrotal oedema *(seek permission)*
Back *(by tilting to a side)*	Percuss/auscultate the posterior chest wall (Pleural effusion) Sacral edema Buttock purpura (HSP) Perianal disease (IBD) Scoliosis (osteodystrophy)
Chest & CVS	Palpate • Rib rosary (osteodystrophy) • Cardiomegaly (CCF, CRF) Auscultate • Flow murmur (CCF) • Gallop rhythm (CCF) • Crepitations (CCF)

STEP IV: GAIT & ANTHROPOMETRY

Squat	To rule out proximal myopathy (steroids)
Check gait	Limp/circumductive gait (stroke)
Must to do	Height , Weight

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) Fundoscopy
4) Detailed motor exam (if relevant)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place. Patient has **generalized edema** i.e. peri orbital puffiness, pedal , sacral , scrotal edema and ascites. His **HR** is ____/min, **R/R** is ___/min. **B.P** is _____mmHg. **Height** and **weight** are at ___th centile.

Apex beat is in left __th ICS in MCL. **Breath sounds** were decreased bilaterally in lower lung fields. There are no signs of **micronutrient deficiency**, any **visceromegaly** or **stigmata of CLD. Gait** is normal.

EDEMA/NEPHROTIC SYNDROME

1) General Look
2) GPE + BP
3) Abdomen, Chest, CNS
4) Gait & Anthropometry

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) **Respiratory Rate for 10 sec**
2) Stature (Short for age?)
3) Nutrition status
4) Face: Periorbital oedema/ Cushingoid features/ sallow complexion
5) Skin: Jaundice, spider naevi, Pallor
6) Skeletal changes of renal osteodystrophy

STEP II: GPE & BP (Sitting on Couch)

Hands & nails	Clubbing (CLD, IBD, CF) Leuconychia (CLD) Palmar crease pallor (CRF) Palmar erythema (CLD)
Wrist	Joint swelling (IBD) Wrist widening (osteodystrophy) **Radial Pulse: rapid** (CCF) **Rate**
BP	**BP** (CRF, various GNs) *(Must to do)*
Arms	Spider naevi (CLD), Muscle bulk: poor (PCM, CRF, CLD) Subcutaneous fat: poor (as above)
Hair	Brittle, sparse (nutritional deficiency)
Neck	JVP: elevated (CCF)
Face	Moon face, hirsute (steroids), Malar flush (SLE causing GN)
Eyes	Peri-orbital edema , Scleral icterus (CLD), Conjunctival pallor (CRF)
Oral cavity	Mouth: uraemic foetor (CRF), Cheilosis, Stomatitis Tongue: Glossitis (nutritional deficiency)
Legs	Pedal edema, striae on thighs
Skin	Rash of kwashiorkor & petechie for vit K deficiency

RECENT ADVANCES

Darbepoetin alfa	Type of EPO : **0.45ug/kg/week** (once a week to once a month)
Erythropoetin beta	Type of EPO (Less painful)
Markers of GFR	Cystatin C Iohexol Plasma Neutrophil gelatinase associated lipocalin Urinary Neutrophil gelatinase associated lipocalin, IL-18, Kidney injury molecule 1
Paricalcitol and **Doxercalciferol**	Newer activated vitamin D analogs

TIPS & TRICKS

1) Measuring B.P. is must in renal cases.
2) A short statured child with pallor/sallow complexion: Think of CKD.

NOTES

kidney transplantation

5. ADJUSTMENT IN DRUG DOSE

1) Lengthening of the interval between doses
2) Decreasing the absolute dose
3) Both

6.DIALYSIS

While waiting for transplant, GFR <15

7.RENAL TRANPLANT

Living donor transplant survival
1yr =90%
2 yr = 85%
5yr = 75%

IV .MONITORING OF DISEASE ACTIVITY

Routine:

1. Hemoglobin levels
2. Serum electrolytes
3. Blood urea nitrogen, creatinine
4. Calcium, phosphorus, , alkaline phosphatase
5. Albumin

Periodic:

1) intact parathyroid hormone (PTH) levels
2) Roentgenographic studies of bone (To detect early evidence of renal osteodystrophy)
3) Echocardiography (To identify left ventricular hypertrophy and cardiac dysfunction)

PROGNOSIS & COUNSELLING

Targets to slow down renal dysfunction

1) Optimal control of hypertension (<95th percentile)
2) Serum phosphorus maintained within the normal range for age and the Calcium–phosphorus product <55 to minimize renal calcium–phosphorus deposition
3) Prompt treatment of infectious complications
4) Prompt treatment of dehydration
5) Correction of anemia with erythropoietin or darbepoetin alfa therapy
6) Control of hyperlipidemia
7) Avoidance of cigarette smoking
8) Prevention of obesity
9) Avoidance of nonsteroidal anti-inflammatory potential nephrotoxic medications
10) Avoid illegal street drugs as well as herbal and or homeopathic medications or "supplements"

Dietary protein restriction not suggested for children because of the concern about adverse effects on growth and development. (Though Suggested for adults)

Oral or intravenous iron supplementation

Resistant to rHuEPO?

1) Evaluate for iron deficiency
2) Occult blood loss
3) Chronic infection
4) Inflammatory state
5) Vitamin B12 or folate deficiency
6) Bone marrow fibrosis related to secondary hyperparathyroidism

Growth retardation / Short stature

Children with CKD : Growth hormone–resistant state

↑ growth hormone levels, ↓ insulin-like growth factor 1 , abnormalities of insulin-like growth factor– binding proteins

Indication for recombinant human growth hormone (rHuGH)

Less than −2 SD for height despite optimal medical support (adequate caloric intake and effective treatment of renal osteodystrophy, anemia, and metabolic acidosis)

Dose of rHuGH : 0.05 mg/kg/24 hr subcutaneously

Duration of treatment

1) Till the patient reaches the 50th percentile for midparental height
2) Achieves a final adult height
3) Undergoes kidney transplantation

3. HYPERTENSION

Cause: Volume overload and/or excessive renin production (glomerular disease)

Hypertensive due to volume overload	Children with proteinuric renal disease
Salt-restricted diet (<2 g/24 hr) CKD stages 1-3: Hydrochlorothiazide 2 mg/kg/24 hr divided bid CKD 4: Furosemide 1-2 mg/kg/dose bid or tid	ACE inhibitors (enalapril, lisinopril) Angiotensin II receptor blockers (losartan) Monitor renal function and electrolyte balance
Uncontrolled hypertension : adjunctive agents	
Calcium channel blockers (amlodipine) β-blockers (propranolol, atenolol) Centrally acting agents (clonidine)	

4. IMMUNIZATIONS

All standard immunizations according to the schedule used for healthy children

Plus yearly influenza vaccine

Exception: withholding live virus vaccines from children with CKD related to glomerulonephritis, or any disease, during treatment with immunosuppressive medications

Must try to administer live virus vaccines for measles, mumps, rubella, and varicella before

2. HORMONAL DEFICIENCIES

Renal osteodystrophy

Goal : To prevent bone deformity and normalize growth velocity

Low-phosphorus diet/Infant formula such as *Similac PM 60/40*

Target phosphorus level

Adolescents	3.5 - 5.5 mg/dL
1-12 yr	4-6 mg/dL

Phosphate binders to enhance GI phosphate excretion

Calcium-based binders (most common)

Non–calcium-based binders such as sevelamer (Renagel) are increasing in use (older children)

Avoid aluminum-based binders : Toxicity

Vitamin D therapy *(The cornerstone of therapy)*

Indication:

1) Patients with 1,25-dihydroxy-vitamin D levels below the established goal range for the child's particular stage of CKD
2) Patients with PTH levels above the established goal range for CKD stage

Calcitriol *(Rocaltrol, 0.25-µg capsules or 1 µg/mL suspension)* **0.01-0.05 µg/kg/24 hr**

Newer activated vitamin D analogs such as **paricalcitol** and **doxercalciferol**

Therapeutic Target: To keep PTH, Ca, PO4 levels within the designated goal range

Target Calcium/phosphorus product (Ca × PO4)

Adolescents	<55 mg^2/dL^2
Younger children	<65 mg^2/dL^2

To minimize the possibility of tissue deposition of calcium phosphorus salts & damage.

Anemia

Cause:

1) Inadequate erythropoietin production (in stages 3–4 CKD)
2) Iron deficiency, folic acid or vitamin B12 deficiency
3) Decreased erythrocyte survival

Recombinant human erythropoietin (rHuEPO) therapy (Decreased the need for transfusion)

Initiated when Hb <10 g/dL

Dose: **50-150 mg/kg/dose subcutaneously 1-3 times weekly** (Target Hb: 11 - 12 g/dL)

*(Alternative to rHuEPO : Darbepoetin alfa (Aranesp) **0.45 µg/kg/wk: Advantage**= once weekly to once monthly dosing)*

		4. Immunizations 5. Adjustment in drug dose 6. Dialysis 7. Renal transplant

b) Details

I. MDT (Multi-disciplinary team approach)

Pediatrician	P. Nephrologist	Endocrinologist	Dietician
Physical therapist	Psychiatrist	Social worker	Nurse clinician

II. NUTRITIONAL MANAGEMENT

Counseling for all children and their families with CKD stages 2 to 5 and 5D.
Patients should receive 100% of estimated energy requirement for age
Tube feeding if oral supplementation is insufficient
Calories balanced between carbohydrate, unsaturated fat and protein.

STAGE	PROTEIN INTAKE
Stage 3 CKD	100-140% of the DRI
Stages 4 and 5 CKD	100-120% of the DRI
Stage 5D	100% of the DRI with allowance for dialysis loss

Children with CKD stages 2-5 should receive 100% of DRI of vitamins and trace elements; water-soluble vitamin supplements are often required in CKD stage 5D.

(Dietary reference intake [DRI])

III. SPECIFIC THERAPY

I. FLUID AND ELECTROLYTE MANAGEMENT

Infants and children (CKD 2° to renal dysplasia): Polyuric + urinary sodium or free water losses = High-volume, low-caloric-density feedings with sodium supplementation.

Children with high blood pressure, edema, or heart failure = Na^+ restriction + diuretic therapy

Fluid restriction only if ESRD requires the initiation of dialysis.

Hyperkalemia
Occurs in

1) Renal function levels when dialysis is initiated
2) Excessive dietary potassium intake
3) Severe acidosis,
4) Hyporeninemic hypoaldosteronism (destruction of the renin-secreting JG apparatus)

Rx
Restriction of dietary potassium intake
Oral alkalinizing agents (Kayexalate)

Acidosis (in almost all children)(Decreased net acid excretion)
Bicitra (1 mEq sodium citrate/mL) or sodium bicarbonate tablets (650 mg =8 mEq of base)
Target serum bicarbonate level >22 mEq/L

	6. MCUG 7. Renal biopsy	7. Radiographs 8. GTT , BSR , HbA1c 9. Serum cholesterol and triglyceride 10. 2 D Echo 11. S.Albumin

b) Details

I. FOR DIAGNOSIS

1. **Inulin clearance** (Gold standard to determine GFR) but it is not easy to measure
2. **Endogenous creatinine clearance** (Most widely used marker of GFR) but creatinine secretion falsely elevates the calculated GFR
   Bedside formula for Children age 1-16 yr (estimates GFR between 15 and 75 mL/min/1.73 m^2) *Estimated GFR = 0.43 × height in cm/serum creatinine in mg/dL*
3. **Cystatin C** (Marker under investigation to determine GFR)
4. **Iohexol** (Marker under investigation to determine GFR)

II. TO FIND ETIOLOGY

1. **Urine analysis** : Hematuria and proteinuria (GN)
   Low specific gravity & minimal abnormalities by dipstick or microscopy (congenital lesions)
2. **Urine Culture**
3. **ANA** (SLE)
4. **Complement levels** (APSGN)
5. **USG KUB** (Anatomical defects)
6. **MCUG** (PUV)
7. **Renal biopsy**

III. TO RULE OUT COMPLICATIONS

1. **CBC** for normochromic, normocytic anemia
2. **BUN & creatinine** (elevated)
3. **ABGs** (pH ↓, HCO_3 ↓)
4. **S. electrolytes** (Hypernatremia, Hyponatremia, Hyperkalemia)
5. **Bone profile** (Hypocalcemia, Hyperphosphatemia, Increased alkaline phosphatase & PTH)
6. **Uric acid** (elevated)
7. **Radiographs** of the hands, wrists, knees (subperiosteal bone resorption + widening of metaphyses)
8. **GTT, BSR, HbA1c**
9. **Serum cholesterol and triglyceride** (elevated)
10. **2 D Echo** (Pericarditis, cardiomyopathy)
11. **S. Albumin** (Hypoalbuminemia)

TREATMENT

Goals of treatment

1) Replace absent or diminished renal functions

a) Tabulated Overview

MDT	Nutrition	Specific R_x
		1. Fluid and electrolyte management 2. Hormonal deficiencies 3. Hypertension

STEP 9: CVS, CHEST & ABDOMEN

CVS: Murmur, Arrythmias
Chest: Rachitic rosary
Abdomen: Mass, bladder, kidney

Redress the child and say thank you!

OFFER (DO IF TIME PERMITS/LONG CASE)

1) Vitals (if Not done)
2) Anthropometry
3) Fundoscopy (Hypertensive changes)
4) Neurological assessment
5) Progressive centile charts

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
He has **tallow complexion** and appears **malnourished** but does not have any **respiratory distress** or **dysmorphism**.
He is **afebrile**; **Pulse** is ___/min, regular in rhythm and normal in volume and character ;
Respiratory Rate is ___/min and **B.P.** _____ mm Hg.
Length is _____cm, **Weight** is _____cm, and **FOC** is _____cm: all below 3rd centile but I would like to plot it on centile chart.
There is no evidence of **proximal myopathy** or **spinal deformity**. **Gait** is normal.
He is **pale**, with **tinge of jaundice** however there is no evidence of **clubbing, leukonychia, palmar erythema, petechiae, bruise, bleed, wrist widening, Scratch marks, joint pain/ swelling, edema, or lymphadenopathy.**
Hearing & Eye examination are normal. **Oral hygiene** is poor with stained teeth.
JVP is not raised. **Thyroid** is not enlarged.

DIFFERENTIALS

1) CKD
2) Celiac disease (short + pale)
3) Rickets (Bony deformities)

INVESTIGATIONS

a) Tabulated Overview

I. FOR DIAGNOSIS	II. ETIOLOGY	III. TO RULE OUT COMPLICATIONS
1. Inulin clearance 2. Creatinine clearance 3. Cystatin C 4. Iohexol	1. Urine analysis 2. Urine Culture 3. ANA 4. Complement levels 5. USG KUB	1. CBC 2. BUN and creatinine 3. ABGs 4. S.electrolytes 5. Bone profile 6. Uric acid

CHRONIC KIDNEY DISEASE

1) General Look
2) Gait + Back
3) Anthropometry
4) Relevant GPE
5) CVS, Chest, Abdomen

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Dysmorphism, Sallow appearance, Nutritional status, Cushingoid face,
2) Glasses, Double lumen catheter, Fistula
3) Respiratory rate

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

1) Squat/Gower's sign
2) Gait
3) Back (Deformity/Tenderness/MMC/sacral edema/Lung bases)

STEP III: ANTHROPOMETRY

Height, Weight, FOC

STEP IV: RELEVANT GPE

Hands	Pallor, clubbing, palmar erythema, Wrist widening
Pulse, **BP*** **	*Avoid BP in fistulous arm*
Arm	BCG scar, Axillary lymph nodes
Scalp	Hair, Anterior fontanelle
Eye	Pallor, Jaundice, aniridia, cataract
Hearing	(Alport syndrome)
Oral cavity	Hygiene
Neck	Thyroid, JVP, CVP site infection, Cervical lymph nodes
Skin	Bruise, bleed
Musculoskeletal	Joint pain/swelling
Leg	Edema

IV. SPECIFIC TREATMENT

Nutritional	Vit D + Nutritional intake of Ca & PO_4 2 strategies for administration of Vit D **Stoss therapy** 300,000-600,000 IU of vitamin D Orally or IM as 2-4 doses over 1 day As doses are observed, it is ideal where adherence is questionable. **Daily, high-dose vitamin D** 2,000-5,000 IU/day over 4-6 wk **Follow up in both strategies** Either strategy should be followed by Daily Vit D intake 400 IU/day if <1 yr | 600 IU/day if >1 yr It is important to ensure adequate dietary calcium and phosphorus; (milk, formula, and other dairy products)
Vitamin D–Dependent Rickets, Type 1	Long-term treatment with 1,25-D (**calcitriol**) Initial doses are **0.25-2 µg/day**, and lower doses are used once rickets has healed. Dose of calcitriol is adjusted to maintain: Low-normal S.Ca | Normal S. PO_4 | High-normal serum PTH
Vitamin D–Dependent Rickets, Type 2	3-6 month trial of high-dose vitamin D and oral calcium. Initial dose of **1,25-D : 2 µg/day** (Some need 50-60 µg/day) **Calcium doses are 1,000-3,000 mg/day** **Patients who do not respond to high-dose vitamin D** may be treated with long-term **intravenous calcium**, with possible transition to very high dose oral calcium supplements. (difficult to treat group)
CKD	**Calcitriol** (permits adequate absorption of calcium and directly suppresses the PTH) **Dietary phosphorus restriction** and the use of **oral phosphate binders** (as hyperphosphatemia is a stimulus for PTH secretion, normalization of the serum phosphorus level is equally important)
XLH, ADHR, ARHR	Oral phosphorus + 1,25-D (calcitriol) PO_4 supplementation is **1-3 g of elemental phosphorus divided into 4-5 doses** Calcitrol : **30-70 ng/kg/day divided into 2 doses**
Hereditary Hypophosphatemic Rickets with Hypercalciuria	Oral phosphorus replacement (1-2.5 g/day of elemental phosphorus in 5 divided oral doses). Treatment of hypophosphatemia decreases serum levels of 1,25-D and corrects the hypercalciuria.

NOTES

OFFER (DO IF TIME PERMITS/LONG CASE)

1) Vitals (if Not done)
2) Anthropometry
3) Developmental assessment

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** BUT I **couldn't gain full cooperation.**
He is **disproportionately short** with **bowing of legs & knock knees.** His **weight** is ____kg & **FOC** is ___cm (I would like to plot it on centile charts)
Heart Rate is ___/min. **Respiratory rate** is___/min and **B.P.** is ____mmHg
There is no **pallor, jaundice, wrist widening, alopecia, frontal bossing, rachitic rosary, Harrison sulcus** and **visceromegaly** .**Spine** is normal. Apart from vitamin D deficiency, I could not appreciate any other **micronutrient deficiency**.

DIFFERENTIALS

YOUNGER CHILD	OLDER CHILD
Nutritional rickets	Vit D dependent rickets
Vit D Resistant rickets	X-linked hypophosphatemic rickets
CKD	CKD

INVESTIGATIONS

Type	Ca++	PO4	PTH	ALP	25 OH Vit D	1,25 OH vit D3	U.Ca++	U.PO4
Nutritional	N or ↓	↓	↑	↑	↓	↓ or N	↓	↑
VDDRI	N or ↓	↓	↑	↑	N	↓	↓	↑
VDDRII	N or ↓	↓	↑	↑	N	↑↑↑	↓	↑
CKD	N or ↓	↑	↑	↑	N	↓	↓	↓
XLH	N	↓	N	↑	N	↓	↓	↑
ADHR	N	↓	N	↑	N	↓	↓	↑
HHRH	N	↓	N	↑	N	↓	↑↑	↑
ARHR	N	↓	N	↑	N	↓	↓	↑

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELING

Depends on cause

III. TREATMENT OF ACUTE PROBLEMS

Symptomatic hypocalcemia

IV calcium bolus : 20 mg/kg of calcium chloride OR 100 mg/kg of calcium gluconate
Continuous intravenous calcium drip
Followed by oral calcium supplements (most infants require 1,000 mg of elemental calcium.), tapered over 2-6 wk in children who receive adequate dietary calcium.

RICKETS/BOWING OF LEGS

1) General Look
2) Gait & Back
3) Anthropometry
4) Relevant GPE

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Dysmorphism, Petechiae & bruises
Respiratory rate

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

1) **Gait** (look for bowing & knock knees)
2) **Back** (Gibbus in Achondroplasia, Kyphoscoliosis in rickets)

STEP III: ANTHROPOMETRY

Height (US: LS) (disproportionately short in skeletal dysplasia & rickets)
Weight, FOC, MUAC

STEP IV: RELEVANT GPE

Hands	Pallor, Clubbing, wrist widening
Pulse	Rate, Rhythm, Volume, Character
B.P.	Increased in CKD
Arm	BCG scar, Axillary lymph nodes
Scalp	Anterior fontanelle, Alopecia (Vit D Dependent Rickets type-2)
Eye	Pallor, Jaundice, Signs of Vit A deficiency
Ears	Deafness (Achondroplasia, Metaphysial dysplasia)
Oral cavity	Stomatitis, Angular cheilosis, Dentition, Bleeding gums
Neck	Thyroid, Cervical lymph nodes
Chest	Rachitic rosary, Pectus carinatum, Harrison sulcus, Auscultate
CVS	Murmur
Abdomen	Hepatosplenomegaly, Inguinal lymph nodes, Scrotum

Redress the child and say thank you!

		Check infections	Full re-assessment
9	**Sensory stimulation** *LMP* *Love* *Mother* *Play*	1) Tender loving care 2) A cheerful, stimulating environment 3) Support for as much maternal involvement as possible (e.g. comforting, feeding, bathing, playing). 4) Structured play therapy for 15–30 min/day 5) Provide suitable toys and play activities for the child 6) Physical activity as soon as the child is well enough	
10	**Prepare for follow-up** Appetite Biotics Complications DEma Social	Transfer to outpatient care/discharge *(A B C DE Social)* Appetite has fully recovered and they are eating well Completed parenteral AntiBiotic treatment, and are clinically well & alert Medical Complications are resolved oeDEma has reduced or resolved **Social** support (Mother/carer's availability, Resources, Counseling of carer on feeding practices) Ask the caregiver to bring the child back for **weekly therapeutic food**, and Ensure the child receives **vaccinations** and routine **vitamin A supplements**	
		Discharge from nutritional treatment Weight-for-height/length ≥ -2 z score + no oedema (for 2 weeks) Mid-upper-arm circumference ≥ 125 mm + no oedema (for 2 weeks) Counsel Mother 1) Five times daily meal (appropriate + correct quantity) 2) High-energy snacks between meals (milk, banana, bread, biscuits) 3) Breastfeed as often as the child wants. 4) Assist and encourage the child to complete each meal. 5) Give food separately to the child (To check child's intake)	
	Follow-up	**Weighed weekly after discharge** (OPD, nutrition rehabilitation centre, local health clinic or health worker) **Refer back to Hospital if:** 1) Failure to gain weight over a 2-week period 2) Loss of weight between two measurements 3) Loss of appetite or oedema	
	Day 7	Give **albendazole** as a single dose or **mebendazole** 100 mg orally twice a day for 3 days (***Started 7 days after admission***)	
	Day 14	**Vitamin A** Last dose Iron at **3 mg/kg per day**	

NOTES

Potassium (mmol)	4	4.2	6.3
Sodium (mmol)	0.6	0.6	1.9
Magnesium (mmol)	0.43	0.46	0.73
Zinc (mg)	2	2	2.3
Copper (mg)	0.25	0.25	0.25

ALTERNATIVE FORMULAS

	F75		F100	
	Dried whole milk based	**Full cream cow's milk**	**Dried whole milk based**	**Full cream cow's milk**
Water: make up to (ml)	1000 ml	1000 ml	1000 ml	1000 ml
Electrolyte/mineral sol	20 ml	20ml	20ml	20 ml
Vegetable oil (g)	20	20	30	20
Milk (g/ml)	**35 g**	**300 ml**	**110 g**	**880 ml**
Sugar (g)	100	100	50	75

II. REHABILITATION PHASE

8

Catch-up growth feeding

Target Calories
150-220 kcal/kg/day

Protein
4-6 g/kg/day

Signs of Rehabilitation Phase **(A H Oedema)**

1) Return of **appetite**
2) No episodes of **hypoglycaemia** (metabolically stable)
3) Reduced or disappearance of all **oedema**

Transition at Hospital

Replace starter F-75 (usually within 1 wk) with an equal amount of catch-up F-100 (100 kcal and 3 g protein per 100 mL) for 2 days.

On the third day if on F-100, **increase each successive feed by 10 ml** until some feed remains uneaten. **(Target 200ml/Kg/Day)**

After the transition phase, refer the child for rehabilitation in outpatient care or to a community feeding programme.

Ready-to-use therapeutic food RUTF	92 g RUTF Packets Containing 500 kcal	Transition Phase **150 kcal/kg/day**	Rehabilitation Phase **200 kcal/kg/day**
	10Kg Child	3 packets/day	4 Packets/day
	First 8 meals per day, and later 5–6 meals per day		

Instructions for mother:

1) Wash hands before giving feeds
2) Sit with the child on the lap and gently offer the feeds.
3) Encourage the child to eat without forced feeding.
4) Offer plenty of clean water in a cup

Monitor for early signs of congestive heart failure

If present Reduce the volume fed to 100 ml/kg per day for 24 h. Then, gradually increase as follows: Day 2 = 115 ml/kg/day , Day 3 = 130 ml/kg/day, Day 4 onwards = 10ml/day

Monitor Rate of weight gain every 3 days as g/kg per day

Good (> 10 g/kg per day)	Moderate (5–10 g/kg/day)	Poor (< 5 g/kg per day)

Calories 100 kcal/kg/day **Protein** 1–1.5 g/kg/day	3) Night feeds are essential 4) If gross edema, reduce volume to 100 ml/kg/day 5) **Feeding tools**: Cup or a bowl, spoon, dropper or syringe 6) As cereal-based F-75 partially replaces sugar with cereal flour, it has the **advantage of lower osmolarity**, which may benefit some children with persistent diarrhoea, but it has to be cooked. 7) Weigh daily and plot weight 8) Keep a 24-hr intake chart. Measure feeds carefully. Record leftovers 9) Therapeutic Combined Mineral Vitamin mix (CMV) contains electrolytes, minerals, and vitamins and is added to ReSoMal and feeds

Days on F-75 (130ml/Kg/Day)	Frequency	Volume/ Kg/Feed	Amount Left	Vomitig	Stool(Frequency/ Consistency)	Daily Wt in grams
1–2	2 h	11ml				
3–5	3 h	16ml				
≥ 6	4 h	22ml				

	F75 (STARTER)	F75 (CEREAL-BASED) (STARTER)	F100 (CATCH-UP)
F75 is also available commercially in which maltodextrins replace some of the sugar and to which potassium, magnesium, minerals, and vitamins are already added.		Lower-osmolality formula : helpful in dysentery/persistent diarrhea	For catch up growth
	No cooking needed	**Cook for 4 min**	No cooking
	Whisk at high speed to prevent oil from separating out.		
Water: make up to (ml)	1000 ml	1000 ml	1000 ml
Electrolyte/mineral sol	20 ml	20ml	20ml
Vegetable oil (g)	30g **(6 tsf)**	30g **(6 tsf)**	60g **(12 tsf)**
Dried skimmed milk (g)	25 **(5 tsf)**	25 **(5tsf)**	80 **(16 tsf)**
Cereal flour (g)	-	35 **(7 tsf)**	-
Sugar (g)	100 **(20 tsf)**	70 **(14 tsf)**	50 **(10 tsf)**
Content per 100 ml			
Energy (kcal)	75	75	100
Protein (g)	0.9	1.1	2.9
% Energy from protein	5%	6%	12%
% Energy from fat	32%	32%	53%
Osmolality (mOsm/L)	413	334	419
Lactose (g)	1.3	1.3	4.2

	3) Add 2l ml of this solution to 1 litre of food to supply the extra potassium and magnesium required 4) Prepare food without added salt (**NO Na^+, NO DIURETICS**)
5 **Infection**	Assume that all children with severe acute malnutrition have an infection **PREVENTION** Minimize risk of cross-infection 1. Avoid overcrowding 2. Wash hands 3. Give measles vaccine to unimmunized children age >6 mo
Antibiotics	**TREATMENT** 1. Ensure all doses are given, and given on time 2. Cover skin lesions so they do not become infected 3. Ensure that there is Gram-negative cover **Note: Avoid steroids as they depress immune function**
	If no complications: **Amoxicillin** PO 25 mg/kg X BD for 5 days If complications (shock, skin lesions ,hypoglycemia, hypothermia, RTI, UTI, or lethargy/sickly): **Ampicillin** (50 mg/kg IV / IM) every 6 hr for 2 days, then PO **amoxicillin** (25-40 mg/kg) every 8 hr for 5 days + **Gentamicin** (7.5 mg/kg IV or IM) OD for 7 days For persistent diarrhea/small bowel overgrowth: Inj **Metronidazole** (7.5 mg/kg X TDS)for 7 days
Anti- M T R	**Anti-Malarials**, **Anti-T.B.**, **Anti-Retroviral Therapy**
Measles Vaccine	≥ 6 months old and not vaccinated **or** vaccinated before 9 months age
Mebenda *	Give **albendazole** as a single dose or **mebendazole** 100 mg orally twice a day for 3 days (***Started 7 days after admission***)
• Anorexia after 7 days of antibiotic treatment, continue for a full 10-day course.	
6 **Micronutrient deficiencies** **AJ's Rule of** **1 FA** **2 Zn** **0.3/3 Cu, Fe** **5 M.Vit, Vit A**	Do not give iron initially, (Wait till second week), because iron can make infections worse **Folic acid at 5 mg on day 1; then 1 mg daily** **Zinc at 2 mg/kg per day** **Copper at 0.3 mg/kg per day** **Iron at 3 mg/kg per day (Day 14)** **Multivitamin syrup at 5 ml** **Vitamin A** on **day 1** and repeat on **days 2** and **14** only if child has any signs of vitamin A deficiency like corneal ulceration or a history of measles < 6 months, 50 000 U (0.6ml/20 drops of A MAX) 6-12 months 100 000 U (1.2ml of A MAX) > 12 months 200 000 U (2.5ml of A MAX)
7 **Initial re-feeding F-75** **Target**	**130 ml/Kg/Day** of F-75 every 2-4 hours (start 2-3 hrly) **100 ml/kg/Day** if child has edema 1) NG feeding if the child is eating < 80% at two consecutive feeds 2) Encourage continued breastfeeding (if breast fed) + F-75

	3) Add 20 ml of this solution to 1 litre of feed to supply the extra potassium and magnesium required. 4) Prepare food without added salt **(NO Na^+ , NO DIURETICS)**
Infection	Assume that all children with severe acute malnutrition have an infection **PREVENTION** Minimize risk of cross-infection 1. Avoid overcrowding 2. Wash hands 3. Give measles vaccine to unimmunized children age >6 mo
Antibiotics	**TREATMENT** 1. Ensure all doses are given, and given on time 2. Cover skin lesions so they do not become infected 3. Ensure that there is Gram-negative cover ***Note:* Avoid steroids as they depress immune function**

If no complications	**Amoxicillin** PO 25 mg/kg X BD for 5 days
If complications (shock, skin lesions ,hypoglycemia, hypothermia, RTI, UTI, or lethargy/sickly)	**Ampicillin** (50 mg/kg IV / IM) every 6 hr for 2 days, then PO **amoxicillin** (25-40 mg/kg) every 8 hr for 5 days + **Gentamicin** (7.5 mg/kg IV or IM) OD for 7 days
For persistent diarrhea/small bowel overgrowth	Inj **Metronidazole** (7.5 mg/kg X TDS)for 7 days

Anti- **M T R**	**Anti-Malarials, Anti-T.B. , Anti-Retroviral** Therapy
Measles Vaccine	≥ 6 months old and not vaccinated **or** vaccinated before 9 months age
Mebenda *	Give **albendazole** as a single dose or **mebendazole** 100 mg orally twice a day for 3 days (***Started 7 days after admission***)
• Anorexia after 7 days of antibiotic treatment, continue for a full 10-day course.	
Micronutrient deficiencies **Aj's Rule of** **1 FA** **2 Zn** **0.3/3 Cu, Fe** **5 M.Vit, Vit A**	Do not give iron initially, (Wait till second week), because iron can make infections worse. **Folic acid at 5 mg on day 1; then 1 mg daily** **Zinc at 2 mg/kg per day** **Copper at 0.3 mg/kg per day** Iron at **3 mg/kg per day (Day 14)** **Multivitamin syrup at 5 ml** **Vitamin A** on **day 1** and repeat on **days 2** and **14** only if child has any signs of vitamin A deficiency like corneal ulceration or a history of measles. < 6 months, 50 000 U (0.6ml/20 drops of A MAX) 6–12 months 100 000 U (1.2ml of A MAX) > 12 months 200 000 U (2.5ml of A MAX)
Initial re-feeding F-75 **Target**	**130 ml/Kg/Day** of F-75 every 2-4 hours… (start 2-3 hrly) **100 ml/kg/Day** if child has edema 1) NG feeding if the child is eating ≤ 80% at two consecutive feeds 2) Encourage continued breastfeeding (if breast fed) + F-75

<95°F (35°C) Rectal temp <95.9°F (35.5°C)	1. Avoid exposure	2. Dress warmly, including head & cover with blanket
	3. Keep room hot; avoid draughts	4. Change wet clothes and bedding
	5. Do not bathe if very ill	6. Feed frequently day and night
	7. Treat infections	

TREATMENT

Actively re-warm

1. **Feed** immediately (Check for hypoglycemia)
2. **Skin-to-skin contact** with carer ("kangaroo technique" : Mother's bare chest/abdomen) or **dress in warmed clothes**, cover head, wrap in warmed blanket and provide indirect heat (e.g. heater; trans warmer mattress; incandescent lamp)
3. **Monitor temperature** hourly (or every 30 min if using heater)
4. Stop re warming when rectal temperature is 36.5°C (97.7°F)
5. **Antibiotics** IV or IM

3

Dehydration

PREVENTION

Replace stool losses: Give ReSoMal after each watery stool. ReSoMal (37.5 mmol Na/L) is a low-sodium rehydration solution for malnutrition

TREATMENT

Do *not* give IV fluids unless the child is in shock

Watery diarrhea + decreased urine output = Some Dehydration

If in Shock: 15 ml/kg over 1 h Ringer's lactate with 5% glucose OR 1/2 Darrow's solution with 5% dextrose OR 1/2 NaCl plus 5% glucose

ReSoMal rehydration PO/NG (Don't give ReSoMal if Cholera is suspected)

First 2 hrs	**5 ml/kg** in 30 min	**5 ml/kg** in 30 min	**5 ml/kg** in 30 min	**5 ml/kg** in 30 min
Next 4-10 hrs With F-75	**5–10 ml/kg/hr** (ReSoMal + F75)	Skip Hour	**5–10 ml/kg/hr** (ReSoMal + F75)	Skip Hour
If rehydration still required at 10 h		**5–10 ml/kg/h F-75 formula**	Skip Hour	**5–10 ml/kg/hr F-75**

Recipe for ReSoMal using standard WHO ORS

2 L water + 1 WHO ORS + Sucrose 50g + 45ml of KCL solution **OR** 40ml electrolyte solution)

Monitoring

Wt. /Vitals/Look (every 30 min for 2 h)(then hourly for the next 4–10 h)

STOP WHEN REHYDRATED (3 or more signs of hydration)

Less lethargic	Improved pulse and respiratory rate	
Eyes less sunken	Tears	Less thirsty
Moist mouth	Skin pinch less slow	Passing urine

STOP IF SIGNS OF OVERLOAD! *RR increasing by 5/min, HR by 25/min, increasing edema; engorged jugular veins*

4

Electrolytes

↑ Na
↓ K, Mg

Deficiencies of potassium and magnesium (take about 2 weeks to correct)

1) Give extra potassium (4 mmol/kg per day)

(Syp K-lyte 40meq/5ml, 10 kg= ½ tsf BD)

2) Give extra magnesium (0.6 mmol/kg per day)

Half-strength Darrow solution with 5% dextrose	Ringer lactate (if all of the above are unavailable)

4. Measure and record **pulse and respirations** at the start and every 10 minutes
If there are signs of improvement (pulse and respiration rates fall)
Repeat IV 15 mL/kg for 1 more hr. Then switch to oral or nasogastric rehydration with ReSoMal, 5-10 mL/kg in alternate hr (see ahead for Rx of dehydration)
If there are no signs of improvement assume septic shock and:
1. **Give maintenance fluid IV (4 mL/kg/hr)** while waiting for blood
2. **Order 10 mL/kg fresh whole blood** and transfuse slowly **over 3 hr.** *If signs of heart failure, give 5-7 mL/kg packed cells rather than whole blood*
3. Give **furosemide 1 mL/kg** IV at the start of the transfusion

HYPOGLYCEMIA : Blood glucose < 3 mmol/L = <54mg/dl : Treat as step 1

SEVERE DEHYDRATION : Do *not* give IV fluids except in shock. Treat as step 3

VERY SEVERE ANEMIA (Hb < 4 g/dL) **OR** (Hb 4-6 g/dL + respiratory distress)
1. Give whole blood 10 mL/kg slowly over 3 hr. If signs of heart failure, give 5-7 mL/kg packed cells rather than whole blood
2. Give furosemide 1 mL/kg IV at the start of the transfusion

EMERGENCY EYE CARE CORNEAL ULCERATION
1. Give vitamin A immediately (age <6 mo 50,000 IU, 6-12 mo 100,000 IU, >12 mo 200,000 IU)
2. Instill 1 drop atropine (1%) into affected eye to relax eye & prevent the lens from pushing out
3. Instil **chloramphenicol or tetracycline & atropine drops** into eye then cover with a **saline-soaked eye pad**, and **bandage** (WHO book)

I.STABILIZATION PHASE

1

Hypoglycaemia
(BSR) < 3 mmol/L OR <54mg/dl

Repeat BSR after 30 min if low **OR** Hypothermic / lethargic

PREVENTION
Avoid long gaps without food and minimize need for glucose:

1. Feed immediately	2. Feed every 3 hr day and night (2 hr if ill)
3. Feed on time	4. Keep warm
5. Treat infections (they compete for glucose)	

Note: Hypoglycemia + hypothermia often coexist : signs of severe infection

TREATMENT
Conscious child:
1) **50 ml of 10% glucose** OR **50ml sucrose solution** (1tsf sugar + 3 Table spoon water) orally or NG OR a feed OR 1 tsf sugar under the tongue
2) Continue 2hr oral/NG Feed for 24 hr (Initially 1/4th of feed every 30min)
3) Keep warm
4) Start broad-spectrum antibiotics

Unconscious Child:
1) **IV 10% glucose at 5 ml/kg** OR through NG
2) No IV/NG: **One tsf of sugar + 2 drops of water** sublingually
3) 2hr NG Feed for 24 hr (Initially 1/4th of feed every 30min) (*look for abdominal distention*)
4) Keep warm
5) Start broad-spectrum antibiotics

2

Hypothermia
Axillary temp

PREVENTION
Keep warm and dry + feed frequently

6) Urine RE
7) Stool RE

TREATMENT

10 STEPS OF TREATMENT

	Stabilization		Rehabilitation
	Day 1-2	Day 3-7	Week 2-6
1.Prevent/Treat hypoglycemia	→		
2.Prevent/Treat hypothermia	→		
3. Treat/ Prevent dehydration	→		
4.Correct electrolyte imbalance	→	→	→
5.Treat Infections	→	→	
6.Correct Micronutrient deficiencies	No iron →	→	With iron →
7.Start cautious feeding	→	→	
8.Rebuild wasted tissue (catch up growth)		→	→
9.Provide loving care and play	→	→	→
10.Prepare for follow up		→	→

Aim of stabilization phase

To repair cellular function, correct fluid and electrolyte imbalance, restore homeostasis, and prevent death from the interlinked triad of hypoglycemia, hypothermia, and infection

Aim of rehabilitation phase : To restore wasted tissues (i.e., catch-up growth)

Practice Essentials

1) Metabolic machinery is repaired before promoting weight gain. Pushing ahead too quickly risks inducing the potentially fatal "refeeding syndrome."
2) Don't treat illness of undernourished child like well-nourished child. It ignores deranged metabolism in malnourished children and can be fatal. It should be considered as severely malnourished with complication (follow 10 steps of R_x)
3) Don't treat edema with a diuretic
4) Don't give a high-protein diet in the early phase of treatment.

Admit if

1. General Danger Signs (Consciousness, Convulsions, Not tolerating orally/Vomiting)
2. Severe Oedema
3. Loss of appetite ± medical complication= complicated SAM

(Good appetite+ No medical complication=Manage in OPD)

Place on Sick list

Isolation & warm environment (25–30 °C)

Emergency Treatment in Severe Malnutrition

SHOCK (Lethargic/unconscious + Cold hands + either:

Slow capillary refill (longer than 3 sec)	**OR**	Weak fast pulse

1. Give **oxygen**
2. Give **sterile 10% glucose (5 mL/kg)** by IV
3. Give **IV fluid at 15 mL/kg over 1 hr**, using:

Ringers lactate with 5% dextrose	Half-normal saline with 5% dextrose

OTHER (DO IF TIME PERMITS/LONG CASE)

1) Vitals (if Not done)
2) Developmental assessment
3) Progressive growth charts
4) Fundoscopy

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___yrs old child who is **conscious** BUT I **couldn't gain full cooperation.** He has a cannula in his right hand. He is **pale, emaciated/with puffy face, apathic look** and **poorly interested** in surroundings with **no distress** or **dysmorphism.**
He is **afebrile; Pulse** is ___/min, regular in rhythm and normal in volume and character ; **Respiratory Rate** is ___/min and **B.P.** _____ mm Hg.
Length is _____cm, **Weight** is _____cm, and **FOC** is _____cm: all below 3rd centile but I would like to plot it on centile chart.
He has **dry shiny/coarse skin; BCG scar** mark is absent/present. **Hairs** are sparse & hypo/ hyper pigmented. There is **cheilosis** and **pedal edema.**
JVP is not raised. **Thyroid** and **genitalia** are normal.
There is no evidence of **rash, clubbing, jaundice, petechiae, bruise, lymphadenopathy, oral ulcers, glossitis, Bitot spot, wrist widening, Harrison sulcus.**
Abdomen is **soft, non-tender** and without any **visceromegaly.**
Chest and **CVS** are unremarkable.
Nappy _____
Back is normal.
He can **hold his neck** but **cannot sit without support.** He can **hold objects** with (palmar/pincer grasp) and **babbles.** His **developmental age** is approximately _____months.

DIFFERENTIALS

MARASMUS	KAWASHIORKER
Primary malnutrition	Primary malnutrition
Celiac disease	Protein losing enteropathy
Chronic infections (T.B./HIV)	CLD
Chronic illnesses (CHD, CLD, CKD)	Nephrotic syndrome

INVESTIGATIONS

I. SUPPORTIVE

1) Hb level (if severe pallor)
2) TLC, DLC (infections)
3) Chest X ray

II. TO ESTABLISH THE CAUSE

1) LFTs, USG Abdomen
2) Anti-TTG Ab
3) 2D-Echo
4) Mantoux test, GeneXpert
5) RFTs & electrolytes, USG KUB

FTT/MALNUTRITION

1) General Look
2) Gait
3) Anthropometry
4) Relevant GPE

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Emaciated/Moon face, Pale looking, Petechiae, bruise, Dehydrated
Respiratory rate

STEP II: GAIT

Ask: *'Bachaa Chal saktaa hai?'*
Check Gait

STEP III: ANTHROPOMETRY

Height, Weight, FOC
MUAC (With arm extended: Measure distance from acromion to olecranon process → Find mid point→ Now flex arm & measure MUAC)

STEP IV: RELEVANT GPE

Hands	Pallor, Wrist widening
Pulse	Rate, Rhythm, volume, character
Arm	Mantoux test mark , BCG scar, Axillary Lymph nodes
Head	Hair, Anterior fontanelle, Frontal bossing
Eye	Pallor, Jaundice, Signs of Vit A deficiency
Oral cavity	Stomatitis, Angular cheilosis, Dentition, Bleeding gums
Neck	Thyroid, Cervical lymph nodes
Chest	Rachitic rosary, Pectus carinatum, Harrison sulcus, Auscultate
CVS	Murmur
Abdomen	Hepatosplenomegaly, Inguinal lymph nodes, Scrotum
Back *(after tilting to side)*	Spinal tenderness, Deformity , Sacral edema, Buttock wasting, Perianal rash

Redress the child and say thank you!

Maintenance therapy of lupus nephritis (any 1 of following)

1) Use of mycophenolate mofetil
2) Every 3 months IV cyclophosphamide
3) Azathioprine for 12 months

III. MONITORING & FOLLOW UP

Anthropometry, BP, BMI, Urine RE, Bone profile, DEXA scan
Reinforce use of Sunscreen + avoidance of prolonged direct sun exposure

PROGNOSIS & COUNSELLING

The severity of disease in pediatric SLE is notably worse than the typical course for most adult-onset SLE.

5 yr survival rate for pediatric SLE	~95%
10 yr survival rate	~80-90%
Mortality: Atherosclerosis & Malignancy	

RECENT ADVANCES

Rituximab	For treatment-resistant glomerulonephritis

NOTES

II. SPECIFIC THERAPY

Drug	Dose	Benefits	Adverse effects
Hydroxy-chloroquine	**5-7 mg/kg/day** Up to 400 mg/day	1) Treats mild SLE manifestations (rash & mild arthritis) 2) Prevents SLE flares 3) Improves lipid profiles 4) Beneficial impact on mortality 5) Beneficial impact on renal outcomes	1) Retinal pigmentation 2) Color vision impairment Ophthalmology exams every 6-12 months
NSAIDs		Management of arthralgias & arthritis	Hepatic, renal,& CVS toxicities
Cortico-steroids	Severe disease IV methyl-prednisolone **30 mg/kg/day** for 3 days, followed by weekly pulses **OR** High doses of oral prednisone **1-2 mg/kg/day**	1) Mainstay R_x for significant manifestations of SLE 2) Improves acute deterioration Limit dose & length of exposure to corticosteroids whenever possible *As disease manifestations improve, corticosteroid dosages are gradually tapered over months*	1) Growth disturbance 2) Weight gain 3) Striae, acne, 4) Hyperglycemia, 5) Hypertension, 6) Cataracts 7) AVN 8) Osteoporosis

Steroid-sparing immunosuppressive agents

Methotrexate	To treat persistent moderate disease, including arthritis, significant cutaneous or hematologic involvement, and pleural disease	
Leflunomide		
Azathioprine		
Cyclophosphamide IV / Oral	For most severe, lifethreatening SLE manifestations, e.g. renal, neurologic, cardiopulmonary	1) Cytopenias 2) Infection 3) Hemorrhagic cystitis 4) Premature gonadal failure 5) Increased risk of future malignancy
Adequate hydration (to attenuate the risk of hemorrhagic cystitis) Use of **gonadotropin-releasing hormone agonists**, such as **leuprolide** acetate (helps prevent gonadal failure)(Fortunately, young girls are at much lower risk of gonadal failure than older women)		
Mycophenolate mofetil		
Belimumab (FDA approved) Monoclonal antibody against BLyS (B-cell activating factor) *(BLyS levels are elevated in SLE relate to disease activity)*	Reduces the number of SLE flares Decreases the dose of prednisone Improves multiple markers of disease severity	Fever Nausea Diarrhea
Rituximab	For treatment-resistant glomerulonephritis?	

The Childhood Arthritis Rheumatology Research Alliance (consensus treatment plan)

Induction therapy of newly diagnosed proliferative lupus nephritis

Glucocorticoid + Cyclophosphamide/mycophenolate mofetil	6 month
Patients failing to achieve a partial response in 6 months it is appropriate to switch agents.	

2) Urine RE for protein
3) Chest X-ray
4) ECG
5) 2D-ECHO

TREATMENT

a) Tabulated Overview

MDT	Supportive R_x	Specific R_x	F/UP
	1. Sunscreen 2. Life style modifications 3. Ca + Vit D 4. Anti-coagulation 5. Immunization 6. Psychiatric consultation 7. Contraception	NSAIDs Corticosteroids Hydroxychloroquine Steroid- sparing Immunosuppressives • Methotrexate • Leflunomide • Cyclopho- sphamide • Mycophenolate mofetil • Belimumab • Rituximab	1. Anthropometry, BP, BMI Urine RE 2. Bone profile 3. DEXA scan 4. Reinforce use of Sunscreen

b) Details

MDT (Multi-disciplinary team approach)

Pediatrician	P. Rheumatologist	Pediatric cardiologist	P. Nephrologist
Psychiatrist	Neurologist	P. Dermatologist	

1. SUPPORTIVE (ALP – OF - TTNM CARE)

Skin	Sunscreen + avoidance of prolonged direct sun exposure/ ultraviolet light
Mucosa	Local applicants
Serosa	1) Attention to cholesterol levels 2) Smoking status 3) Body mass index, blood pressure 4) Statins (pubertal patients with an elevated CRP)
Joints	Adequate intake of calcium and vitamin D (to prevent future osteoporosis)
Kidneys	-
Antibodies	Antiphospholipid antibody syndrome: Long-term anticoagulation to prevent thrombotic events
Blood	1) Routine immunization 2) Annual influenza vaccination 3) 23-valent pneumococcal vaccine 4) Prompt attention to febrile episodes (evaluation for serious infections)
CNS	Psychiatric consultation

Contraception

Counsel adolescent girls about these risks of pregnancy & appropriate contraceptive options.

1) Pregnancy can worsen SLE
2) Obstetric complications are more common in SLE.
3) Many of the medications used to treat SLE are teratogenic

INVESTIGATIONS

a) Tabulated Overview

I. FOR DIAGNOSIS	II. SUPPORTIVE	III. TO RULE OUT COMPLICATIONS
1. ANA 2. Anti–ds DNA 3. Anti-Smith 4. Hypo-complementemia 5. Anti-phospholipid Ab 6. CBC 7. Coomb's test	1. ESR 2. CRP 3. Hypergammaglobulinemia 4. Antiribonucleoprotein Ab 5. Anti-Ro & La antibodies 6. Antihistone antibodies	1. RFTs & electrolytes 2. Urine RE for protein 3. Chest X-ray 4. ECG 5. 2D-ECHO

b) Details

I. FOR DIAGNOSIS

Test	Sensitivity	Specificity	Comments
ANA	95-99%	~50%	ANA-negative lupus is extremely rare Up to 20% of healthy individuals also have a positive ANA test result (poor screening test) Not reflective of disease activity
Anti–ds DNA	40-65%	~98%	More specific, correlate with disease activity (esp in Nephritis)
Anti-Smith	40-65%	~98%	More specific, does not correlate with disease activity
Hypo-complementemia	Decreased in active disease: Not represented among the ACR classification		
Anti-phospholipid Ab	Increase clotting risk, can be found in up to 66% of children and adolescents		
CBC	Anemia, Leukopenia, Lymphopenia,Thrombocytopenia		
Coomb's test	Positive direct Coombs test (in the absence of hemolytic anemia)		

II. SUPPORTIVE

ESR	Often elevated in active disease
CRP	Correlates less well with disease activity Acutely elevated CRP = Infection Chronic mild elevation of CRP = Increased cardiovascular risk
Hypergammaglobulinemia	common but nonspecific finding
Antiribonucleoprotein antibody	Increased risk for Raynaud phenomenon & pulmonary HTN High titer may suggest mixed connective tissue disorder
Anti-Ro antibody (anti-SSA antibody) Anti-La antibody (anti-SSB antibody)	**Associations** Sicca syndrome — Sjögren syndrome Increased risk of neonatal lupus in offspring (congenital heart block) Cutaneous and pulmonary manifestations of SLE Isolated discoid lupus
Antihistone antibodies	Drug-induced lupus, May be present in SLE

III. TO RULE OUT COMPLICATIONS

1) RFTs & electrolytes

SYSTEMIC LUPUS ERYTHEMATOSUS

1) Inspection
2) Palpation
3) Gait + Back
4) Range of motion
5) Relevant GPE
6) Functional ability
7) Measurements

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient [Lying/Sitting on couch/parent's lap]**
d. **Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)**
e. **Right side to approach for examination**

STEPS OF EXAMINATION

See Locomotor system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

See Locomotor system exam for details

DESCRIPTION

See Locomotor system exam for details

DIFFERENTIALS

Infections	Sepsis, Epstein-Barr virus, parvovirus B19, endocarditis
GN	Poststreptococcal glomerulonephritis
Rheumatologic	sJIA, vasculitides
Malignancies	Leukemia , Lymphoma
Drugs	**Antibiotics**: Minocycline, Sulfonamides, tetracycline, penicillin **Anti-TB** : Isoniazid, Rifampin **Anti-hypertensive**: Hydralazine , diltiazem, methyldopa, beta blockers, captopril **Anticonvulsants**: chlorpromazine, Phenytoin, ethosuximide, carbamazepine, valproate **Anti-arrhythmic agents** : Procainamide, amiodarone, quinidine **Chelators & heavy metals**: Penicillamine, lithium, gold **Biological agents**: interferon- , etanercept, infliximab, adalimumab

RECENT ADVANCES

Oral Janus kinase (JAK) inhibitors	Used in Systemic JIA e.g. Tofacitinib, Ruxolitinib Inhibit JAK signaling pathways involved in immune activation &inflammation

NOTES

	Treat infection Corticosteroids, Etoposide, intrathecal Methotrexate Antithymocyte globulin (ATG) and Cyclosporine for maintenance therapy **Allogeneic stem cell transplantation**
Psychosocial adaptation	Disability not directly associated with arthritis (20% of patients) Chronic pain syndromes (20% of patients) Rx **Counseling by mental health professionals**

V. FOLLOW UP/MONITORING OF DISEASE ACTIVITY

1) **Clinical monitoring** : Fever, Anemia, joint swelling/decreased ROM, Constitutional symptoms (appetite, fatigue)
2) **Periodic Slit lamp exam**

High risk : 3monthly	Moderate risk: 6monthly	Low risk : Yearly
Poly/oligo articular ANA+ Age of onset : ≤ 6yr old Duration of disease : ≤ 4yr	Poly/oligo articular ANA+/- Age of onset : > 6yr old Duration of disease : 4-7yr	Systemic disease ANA -ive Age of onset : > 6yr old Duration of disease : > 7yr

3) **Laboratory parameters** : Hb, WBCs, Plt, ESR, CRP, S.Albumin (↑), S. Globulin (↓)
4) **Imaging parameters** : X-ray of joints

PROGNOSIS & COUNSELLING

Oligoarticular without uveitis : Best outcome	Polyarticular (RF+)/sJIA : Poorer prognosis

Without TNF-α inhibitors: 50% have active disease persisting into early adulthood – severe limitations of physical function.

OLIGOARTICULAR JIA

Extended oligoarticular disease : Poorer prognosis

Under 6 yr old Girls having Oligoarthritis + ANA-positive = Greatest risk for chronic uveitis ⇒ posterior synechiae, cataracts, glaucoma, and band keratopathy, with resultant blindness

No association between the activity or severity of arthritis and uveitis.

POLYARTICULAR JIA

More prolonged course of active joint inflammation and requires early and aggressive therapy.

Predictors of severe and persistent disease :

Young age at onset	RF seropositivity or rheumatoid nodules
Large numbers of joints	Disease involving the hip and hand and wrist

SYSTEMIC JIA (most difficult to control)

Poorer prognosis:

Polyarticular distribution of arthritis	Fever lasting >3 months
Increased inflammatory markers, such as platelet count and ESR, for >6 months	

Types

Persistent (55%)	**Polycyclic/recurrent**(34%)	**Monocyclic** (11%)
> 33% will have permanent disability with active disease in adult life.		Monocyclic do well

Death

Infection	MAS	Amylodosis

	Canakinumab (IL-1β inhibitor) (FDA-approved: sJIA in > 2 yr old) Tocilizumab (IL-6 receptor antagonist) (FDA-approved: sJIA in > 2 yr) 3) **MTX and anti-TNF agents** (Less responsive) 4) Oral **Janus kinase (JAK) inhibitors** (tofacitinib, ruxolitinib) inhibit JAK signaling pathways involved in immune activation and inflammation.

IV. TREATMENT OF COMPLICATIONS

ARTICULAR	EXTRA-ARTICULAR
1) Joint contractures 2) Joint deformities 3) Leg length discrepancy 4) Popliteal cysts	1) Infections 2) Anemia 3) Growth failure 4) Psychosocial adaptation 5) Uveitis 6) Pleural effusion 7) Myocarditis 8) Gastritis 9) MAS 10) Amylodosis

ARTICULAR

Joint contractures & deformities	Flexion contractures of the knees, hips, and wrists **R_x** 1) Aggressive medical control of arthritis 2) Intraarticular corticosteroid injections 3) Appropriate splinting 4) Stretching of the affected tendons
Leg length discrepancy	Shoe lift on the shorter side to prevent secondary scoliosis
Popliteal cysts	1) No treatment if they are small 2) Intraarticular corticosteroid injection in the anterior knee

EXTRA-ARTICULAR

Infections	IV antibiotics
Anemia & Growth failure	**Causes:** Nutritional | Anemia of Chronic disease | GI bleed due to NSAIDs sJIA | Bone marrow suppression | Megaloblastic anemia **R_x** : Iron, Folic acid, Dietary therapy
Uveitis	*Periodic slit-lamp ophthalmologic examinations for asymptomatic uveitis* **Initial** : Mydriatics + corticosteroids (topically, systemically, or through periocular injection) **Severe uveitis** : Methotrexate and antibodies to TNF-α (adalimumab and infliximab)
Pleural effusion	Chest intubation
Gastritis	H_2 receptor antagonists
MAS	**Emergency treatment** : High-dose IV methylprednisolone, cyclosporine, or anakinra (IL-1 inhibitor) **Severe cases** : Therapy similar to that for primary HLH

Slit lamp exam	To rule out Uveitis
Synovial fluid	RE & Biopsy: To rule out Infective complications/origin
BMA	To rule out HLH/Malignancy

TREATMENT

a) Tabulated Overview

MDT	Supportive R_x	Specific R_x	R_x of Complications	F/UP
As Below	1) Education 2) Pain relief 3) Diet 4) Physiotherapy 5) Occupational therapy	1) NSAIDs 2) DMARDs 3) Steroids 4) Biologic agents	1) Articular 2) Extra-articular	1) Clinical 2) Slit lamp exam 3) Lab 4) Imaging

b) Details

I. MDT (Multi-disciplinary team approach)

Pediatrician	Rheumatologist	Ophthalmologist	Orthopedic surgeon
Physical therapist	Dietician	Social worker	Nurse clinician
Psychologist	Psychiatrist		

II. SUPPORTIVE TREATMENT

Education	Parent & child
Pain relief	Paracetamol, Nocturnal NSAIDs, Splinting
Diet	Dietary evaluation & counseling Appropriate calcium, vitamin D, protein, and caloric intake
Physiotherapy	To optimize joint function
Occupational therapy	A social worker and nurse clinician can be important resources

III. SPECIFIC THERAPY

Polyarthritis (RF -ve)	**Standard therapy** : MTX + NSAIDs (MTX takes 6-12 wk to affect) **Nonresponsive** : Anti-TNF agents (e.g., etanercept, adalimumab) Biologics (abatacept)
Polyarthritis (RF +ve)	**MTX + Anti-TNF agents** (Early aggressive therapy as Long-term remission unlikely)
Oligoarthritis Psoriatic arthritis Enthesitis-related arthritis	1) **NSAIDs** (First line) 2) **Intraarticular steroids** (Triamcinolone hexacetonide) (If no/partial response after 4-6 wk of NSAIDs or functional limitations) 3) **MTX** (minority of patients) **Sulfasalazine** (alternative in **ERA**) 4) **TNF inhibitors** (Not responsive to MTX)
Systemic arthritis	1) **Systemic steroids** For severe systemic illness / For control of uveitis Bridge therapy during the wait for therapeutic response to a DMARD (Steroids do not prevent joint destruction) 2) **IL-1 or IL-6 inhibitors** (dramatic and rapid response) Anakinra(For severe disease) (IL-1 inhibitor)

	3) Leukemia 4) Neuroblastoma 5) Bone tumor (osteosarcoma, Ewing sarcoma)

INVESTIGATIONS

a) Tabulated Overview

I. FOR DIAGNOSIS	II. SUPPORTIVE	III. TO RULE OUT COMPLICATIONS
Clinical diagnosis	1) **CBC** : Hb, WBC, Platelets 2) **Serology** : ESR, CRP, Ferritin, ANA, RF, HLA-B27, Immunoglobulins	1) **Radiology** : Joint, Chest X ray 2) **ECG** 3) **2D Echo** 4) **Slit lamp examination** 5) **Synovial fluid** : RE, Biopsy 6) **Bone marrow aspiration/biopsy**

b) Details

I. FOR DIAGNOSIS

Clinical diagnosis

II. SUPPORTIVE

CBC	1) **Microcytic anemia** (Systemic, Polyarthritis, Psoriatic) (Hb 7-10 g/dL) 2) **WBC** ↑↑ (Systemic) 3) **Platelets** ↑↑in Systemic (normal or ↓ in MAS) ** Criteria of MAS : (↓N) WBC count and/or platelet count + active sJIA*
Serology	1) **ESR** ↑↑ (All types) (↓ in MAS) 2) **CRP** ↑↑ (All types) 3) **Ferritin** ↑ (Systemic) (Markedly raised in MAS >10,000 ng/mL) 4) **ANA positive** (60%= Oligoarthritis, 50% = Psoriatic, 40% = Polyarthritis:RF-negative) rare with sJIA associated with increased risk of chronic uveitis 5) **RF positive** (Polyarthritis: RF-positive) 6) **HLA-B27 positive** (80% of Enthesitis-related) 7) **Immunoglobulin levels** ↑ (systemic) 8) **Anti–cyclic citrullinated peptide antibodies** (anti-CCP Ab) *ANA &RF seropositivity can occur with transient events(e.g. viral infection)*

III. TO RULE OUT COMPLICATIONS

Radiology /MRI	1) **Joint (X-ray, MRI*)** Early radiographic changes (MRI more sensitive) Soft tissue swelling | Periarticular osteopenia | Periosteal new-bone apposition Continued active disease Subchondral erosions | Loss of cartilage | Bony destruction, and fusion Changes in neural arch joints(C2-C3) may progress to atlantoaxial subluxation 2) **Chest X-ray** : To rule out pleural effusion
ECG & Echo	To Rule out cardiac complications

JUVENILE IDIOPATHIC ARTHRITIS

1) Inspection
2) Palpation
3) Gait + Back
4) Range of motion
5) Relevant GPE
6) Functional ability
7) Measurements

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

See Locomotor system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

See Locomotor system exam for details

DESCRIPTION

See Locomotor system exam for details

DIFFERENTIALS (*Mnemonic: Joints ache* ***In TrAM***)	
Infections	1) Septic arthritis 2) Osteomyelitis 3) Reactive arthritis 4) Tuberculous arthritis 5) Acute rheumatic fever
Trauma	1) Local trauma 2) Avascular necrosis (Legg-Calvé-Perthes disease) 3) Slipped capital femoral epiphysis
Autoimmune	1) JIA 2) SLE 3) JDM/MCTD/Scleroderma 4) IBD 5) Autoimmune hepatitis
Malignancy & Hematology	1) Hemophilia 2) Sickle cell disease

weekly or monthly IV dosing along with daily oral corticosteroids as needed.
Monitoring parameters
Muscle enzymes normalize
Muscle strength improves

METHOTREXATE (steroid-sparing agent)
1 mg/kg or 15 mg/m^2, (maximum 40 mg)
Weekly oral, intravenous, or subcutaneous
The concomitant use = Methotrexate + steroid (halves the cumulative dosage of steroids)
Folic acid 1 mg daily to reduce toxicity and side effects of folate inhibition (oral ulcers, nausea, and anemia)
CAUTION
Avoid live-virus vaccination, although inactivated influenza vaccination is recommended yearly.

DISEASE-MODIFYING AGENT - Hydroxychloroquine
Little toxicity risk
4 - 6 mg/kg/day orally

BIOLOGICAL AGENTS – RITUXIMAB (Recent advancements)
Unresponsive disease
Steroid-dependent patients Resistant inflammatory myopathies

OTHER MEDICATIONS for severe unresponsive disease include
1) IV immunoglobulin
2) Mycophenolate mofetil
3) Cyclosporine
4) Cyclophosphamide

NOTES

4. **Muscle biopsy** (non diagnostic in 20%)
Necrosis Inflammation

II. SUPPORTIVE

1. **CBC** (anemia consistent with chronic disease)
2. **ESR** (normal)
3. **ANA** + (>80%)
4. **Antibodies to SSA, SSB, Sm, ribonucleoprotein, and doublestranded DNA** (negative)
5. **Myositis-specific autoantibodies** +
i.Antibodies to Pm/Scl (for myopathies with pulmonary interstitial fibrosis and/or cardiac involvement)
ii.Anti–Jo-1, anti–Mi-2, anti-p155/140, anti-NXP2
6. **Rheumatoid factor test** = negative
7. **NCS** (normal)
8. **A contrast swallow study** to document palatal dysfunction and risk of aspiration
9. **Pulmonary function testing** for restrictive defect and vital capacity
10. **Radiography for Calcinosis**

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELING

The mortality rate 1%
Active symptoms persist <1.5 yr
At 7 yr of follow-up
75% of patients have little to no residual disability
25% continue to have chronic weakness
40% have chronic rash

Up to one-third may need long-term medications to control their disease.

III. SUPPORTIVE TREATMENT

1) **Nasogastric or gastrostomy feedings** to avoid aspiration (pharyngeal weakness)
2) **Full bowel rest** (GI vasculitis)
3) **Ventilator therapy / tracheostomy** (Severe respiratory weakness)
4) **Physical therapy and occupational therapy**
Bed rest is not indicated, because weight bearing improves bone density and prevents contractures.
5) **Social work and psychology services**
6) **Avoid sun exposure and apply high sun protection factor sunscreen daily, even in winter and on cloudy days.**
7) **Vitamin D and calcium supplements** are indicated for all children undergoing long-term corticosteroid therapy, in an attempt to reduce osteopenia and osteoporosis from medication.

IV. SPECIFIC TREATMENT

CORTICOSTEROIDS (mainstay of treatment)

Oral prednisone at 2 mg/kg/ day (maximum 60 mg daily) tapered over a period of 12 mo (IV in Children with GI involvement)
In more-severe cases with respiratory or oropharyngeal weakness, high-dose pulse **methylprednisolone** is used (30 mg/kg/day for 3 days, maximum dose 1 g/day) with ongoing

JUVENILE DERMATOMYOSITIS

1) Inspection
2) Palpation
3) Gait + Back
4) Range of motion
5) Relevant GPE
6) Functional ability
7) Measurements

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

See Locomotor system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

See Locomotor system exam for details

DESCRIPTION

See Locomotor system exam for details

DIFFERENTIALS

1) JDM
2) Juvenile polymyositis
3) SLE
4) MCTD

INVESTIGATIONS

I. FOR DIAGNOSIS

1. **Muscle-derived enzymes** (creatine kinase, aldolase, aspartate aminotransferase, alanine aminotransferase, and lactate dehydrogenase)
Most commonly elevated on initial presentation = Alanine aminotransferase
2. **EMG** (Muscle weakness)
3. **MRI using T2-weighted images and fat suppression**
Identifies active sites of disease, increasing the sensitivity of muscle biopsy and EMG

IV. SPECIFIC TREATMENT

Steroids

Prednisone (0.75 mg/kg/day) for the 1st 10 days of each month to avoid chronic complications.
Deflazacort 0.9 mg/kg/day, may be more effective than prednisone.
Fluorinated steroids, such as dexamethasone or triamcinolone, should be avoided because they induce myopathy by altering the myotube abundance of ceramide.

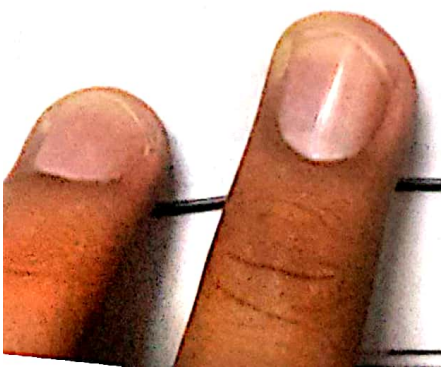
V. RECENT ADVANCEMENTS

I/V or S/C injection of **antisense oligonucleotide drugs** (**Drisapersen** and **eteplirsen**) that induce exon skipping during mRNA splicing in patients with susceptible mutations (~15% of patients) to restore the open reading frame in the DMD gene.

TIPS & TRICKS

1) Remember to examine heart in cases of muscle problems.

NOTES



2. Electromyography (EMG) shows characteristic myopathic features but is not specific for DMD. No evidence of denervation. Motor and sensory nerve conduction velocities are normal.

3. TO RULE OUT COMPLICATIONS

1. Cardiac assessment by **echocardiography, electrocardiography (ECG), and radiography of the chest** is essential and should be repeated periodically.

TREATMENT FOR DMD

I. MDT (Multi-disciplinary team approach)

Cardiologist, psychologist, physiotherapist, orthopedic surgeon
Beware of malignant hyperthermia after anesthesia

II. PARENTAL COUNSELING

Genetic counselling
Death occurs usually at about 18-20 yr of age.
The causes of death

1) Respiratory failure during sleep
2) Intractable heart failure
3) Pneumonia,
4) Aspiration
5) Airway obstruction

III. SUPPORTIVE TREATMENT

Cardiac decompensation
Refer to a pediatric cardiologist
Often responds initially well to digoxin

Pulmonary infections
Prompt treatment
Avoid contact with children who respiratory / contagious illnesses
Immunizations for influenza virus and other routine vaccinations

Preservation of a good nutritional state
Avoid excessive doses of vitamins (not a vitamin-deficiency disease)
Adequate calcium intake (To minimize osteoporosis in boys confined to a wheelchair)
Fluoride supplements (if the local drinking water is not fluoridated)
Obesity (Sedentary children burn fewer calories + depression & eat excessively) Dietary restrictions with supervision may be needed.

Physiotherapy (Delays but does not always prevent contractures)
Contributes little to muscle strengthening because patients are already using their entire reserve for daily function, and exercise cannot further strengthen involved muscles. Excessive exercise can actually accelerate the process of muscle fiber degeneration.

Surgical correction of the elbow contracture
Technically feasible, but the result may be deleterious.

Watch for progressive scoliosis
Treated early by orthopedists using external braces or corsets and occasionally by surgeons.

2) Anthropometry
3) Functional assessment
4) Developmental assessment

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____ (Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
There is no **distress** or **dysmorphism**.
His has **waddling gait** with **lordotic posture**; **calf hypertrophy** and **positive Gower sign** but no evidence of **winging of scapula**.
He has **proportional bulk** in all four limbs, **normal tone, power of 4/5 in proximal group of muscles** and **5/5 in distal muscle groups**.
He has **diminished reflexes** and **downgoing plantars**. There is **no ankle clonus**. **Superficial reflexes** are intact. Upper limbs are not showing any evidence of **hypertrophy at fore arm**.
Spine is lordotic at lumber level.
All accessible **cranial nerves** are intact.
Apex beat is located at _____ICS lateral to MCL with normal **heart sounds**.
There is no evidence of **aspiration** or **tongue fasciculation**.

DIFFERENTIALS

1) **Duchenne muscular dystrophy**
2) **Becker muscular dystrophy**
3) **Limb girdle muscular dystrophy**
4) **SMA-3**

INVESTIGATIONS FOR DMD

1. FOR DIAGNOSIS

1. **Polymerase chain reaction (PCR)** for the dystrophin gene mutation (Primary test)

2. **Clinical features + Raised serum CK** (15,000-35,000 IU/L (normal <160 IU/L)) greatly elevated in DMD, even in presymptomatic stages, including at birth. A normal serum CK level is incompatible with the diagnosis of DMD (Terminal stages CK value may be considerably lower because there is less muscle to degenerate)

3. **Muscle biopsy** done if PCR is normal and clinical suspicion is high

4. **Dystrophin immunocytochemistry** (More specific) performed on muscle biopsy sections detects the 30% of cases that do not show a PCR abnormality.

5. **Dystroglycans and other sarcolemmal regional proteins**, such as merosin and sarcoglycans, also can be measured because they may be secondarily decreased.

2. TO FIND ETIOLOGY/SUPPORTIVE

1. **Other lysosomal enzymes** present in muscle, such as aldolase and aspartate aminotransferase, are also increased but are less specific.

DUCHENNE MUSCULAR DYSTROPHY

1) General Look
2) Gait + Back
3) Motor system
4) Relevant GPE
5) CVS, Chest

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building (Talk to child; note voice)**
c. **Position patient [Lying/Sitting on couch/parent's lap]**
d. **Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)**
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) **Look for myopathic facies**
2) **Mood, intelligence, ability to communicate**
3) Describe the **resting posture/ standing posture**
4) **Wheelchair** (describe child's posture + Appropriateness of chair)
5) **Respiratory rate** (LRTI, Heart failure)

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

1) **Inspect legs & upper thigh** (calf hypertrophy, thigh wasting, scars, pes cavus)
2) **Inspect back for spinal deformities** (Lordosis, Scoliosis, Kyphosis)
3) **Gower sign** (Sit down & stand without support of ground)
4) **Ask to walk** (waddling gait, lordotic posture)
5) **Ask to press with both hands against wall** (for scapular winging)

STEP III: MOTOR SYSTEM

See Motor system exam for details

STEP IV: RELEVANT GPE

Oral cavity	For tongue fasciculation (SMA), Hypertrophy (DMD)
CVS	For DCMP, Pulmonary hypertension
Chest	Aspiration

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)

STEP VI: FUNCTIONAL ABILITY

Comb your hair	*Write your name with pen*

STEP VII: MEASUREMENTS

True leg length	Anterior superior iliac spine to medial malleolus
Apparent leg length	Umbilicus to medial malleolus

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Missed Steps
2) Vitals (if Not done)
3) Anthropometry
4) Slit lamp examination of eyes

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
He has no **distress** or **dysmorphism**.
Gait is **normal/antalgic** and examination of **back** is unremarkable.
He has 6 **swollen joints** including Right knee and elbow **with/without signs of active inflammation** (in form of redness, warmth and tenderness)
Range of motion is reduced during extension/flexion at Right knee.
He has no **fixed flexion deformity** or **contracture**.
He is **afebrile** (to touch). **BCG scar** is present.
There is no evidence of **steroid toxicity, pallor, nail pitting, psoriasis, malar/discoid rash, oral/nasal ulcers, purpura, visceromegaly, lymphadenopathy** or **pericardial rub**.
He can **dress/undress himself & able/not able to walk.**
His **true leg length** for both sides is equal i.e. ____cm each
His **apparent leg length** for both sides is equal i.e. ____cm each.

TIPS & TRICKS

1) It is a time taking examination and needs practice time and again. Don't worry if you miss few steps; you can offer them.
2) **Never Ever inflict pain to child** while examining a tender local joint. You can tell examiners if there are limitations in examining a specific joint.
3) Always look towards the face of child in locomotor examination.
4) Expose properly because the child can have purpura or bruises.
5) Any movement which turns the anterior surface of the limb laterally is external rotation and any movement which turns the anterior surface of limb medially is internal rotation.
6) Do passive movements only in affected joints.

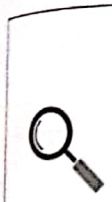
NOTES

Wrist	**Reverse prayer sign** (Wrist flexion) **Deviation at wrist**: Do radial then ulnar deviation
Elbow	**Pose to Turn key of car** (with elbow fixed at flexion) **Supination , Pronation** **Flexion at elbow** **Extension at elbow**
Shoulder	**Embrace yourself** (adduction) **Place both hands behind occiput** (Flexion & abduction) **Scratch your back** (External rotation) **Stretch your arms outward & make circular movements** (Circumduction) **Place your hands on back** (Internal rotation & extension)
Neck	**Touch chin with chest** (Flexion) **Touch occiput with back** (Extension) **Rotate neck right & left** *('Slaam phairaain')* (Rotation) **Touch ear with shoulder** (Lateral flexion)
TM Joint	**Open your mouth and place three fingers in mouth**
Spine	**Sway on both sides with arms on pelvis** (Rotation)
ASK CHILD TO LAY DOWN ON COUCH	
Feet	**Dorsi & plantar flexion** **Inversion & eversion** **Check for tenderness of small joints**
Knee	**Flexion, Extension,** **Internal & external rotation**
Hip	**Abduction, Adduction** **Medial & lateral rotation** **Flexion, Extension** (by turning to one side or lying prone)
SI joint	**Check for tenderness** by pressing at both pelvic areas with opposite hands

STEP V: RELEVANT GPE (Child lying on couch)

Hands & nails	Rash, Pitting of nails, Digital ulcer, deformity of finger (boutannier/ swan neck), muscle wasting, gottrons papules over PIP&MCP joints
Pulse	Rate, rhythm , Volume, character
Elbow	Rash, Sub cutaneous nodule
BCG scar	Check for presence
Hair	Alopecia (SLE)
Face	Look for micrognathia, Butterfly rash (SLE)
Eye	Pallor, Redness Examine with pen torch –Pulpillary size, photophopia, synachae, cataract
Nose	Nasal ulcers (SLE)
Oral cavity	Ulcers (SLE)
Thyroid	For autoimmune thyroiditis
CVS	Murmur/Pericardial rub
Abdomen	Hepatosplenomegaly (soJIA, SLE, Malignancy)
Genitalia	Seek permission for tanner stage & mass, **Inguinal Lymph nodes**
Make child sit on couch for **Cervical & axillary lymph nodes**	

LOCOMOTOR EXAMINATION



1) Inspection
2) Palpation
3) Gait + Back
4) Range of motion
5) Relevant GPE
6) Functional ability
7) Measurements

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient [Standing in front of you]**
d. **Exposure: Ask child to remove his shirt by himself** (*To check functional ability*) (Ideal exposure in male child: Shirt off, trousers rolled up) (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: INSPECTION (Make child stand in front of you; adequately exposed)

1) **Dysmorphism**
2) **Joints** (Symmetry/swelling/loss of normal countor/ deformity/ redness / muscle wasting)
3) **Skin**: Rash, bruises, BCG scar, Signs of steroid toxicity (Acne, moon face, hirsutism, striae)

STEP II: PALPATION OF JOINTS

Ask: *Kissi Jorr main dard tuu nahi?* (Examine affected joint in the end!)
1) **Temperature** (Compare bilaterally with dorsum of hand)
2) **Tenderness** (while looking into eyes of child)
3) **Fluctuation**

STEP III: GAIT & BACK

Back: Check Range of motion at spine

Touch toes	Flexion (see from side & back), check for spinal tenderness/ deformity
Arch back	Extension
Lateral bending (both sides) with hands on pelvis	

Ask: *'Bachaa Chal saktaa hai?'*
1) **Squat** (for proximal weakness)
2) **Stand on one leg :** look at sacral area for Trendelenburg sign
3) **Ask to walk** (Antalgic gait)

STEP IV: RANGE OF MOTION (Make child sit on couch: Stand in front to demonstrate)

Fingers	**Make fist with thumb inside** (Observe is it closing fully? ROM? Tenderness?) **Approximate each finger with thumb** (counting on digits) *(Tasbeeh)*
Hands &	**Prayer sign** (Wrist extension)

III. SPECIFIC TREATMENT

NEUROBLASTOMA	WILM'S TUMOR
Stage I & II : Surgery **Stage III & IV:** Chemotherapy	**Stage I & II** : Chemotherapy (Actinomycin, Vincristine, Doxorubicin) **Stage III** : Chemotherapy + Radiation **Stage IV & V**: Chemotherapy + Nephrectomy + Radiation

TIPS & TRICKS

1) Usually the difficult cases like it have more chances to pass if you follow basics in that case.

NOTES

OFEER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals
2) Anthrpometric measurements
3) Fundoscopy (for hemorrhage)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
He has **pallor** and **abdominal distension** but no **distress** or **dysmorphism.**
He is **afebrile** (to touch) with **Respiratory Rate** = ___ /min, **BP** ____mmHg
Pulse is ___/min regular in rhythm and normal in volume & character
There are no **scar, striae, rash, petechiae** or **scratch marks.**
Abdomen is **soft; non-tender** with a **7 x 8 cm** swelling noted at **Right lumbar region** which is **crossing/not crossing midline.**
Overlying skin is of normal color, temperature & texture with/without discharge/sinus.
Swelling is **non-tender/ tender** with **rounded margins, smooth / irregular surface & soft / firm / hard consistency.**
It is **non-mobile / mobile & attached** to Skin/Underlying Structures
No **pulsations / thrill or bruit** is noted.
There is no evidence of **pallor,bruises, petechiae, hepatosplenomegaly , lymphadenopathy, bone tenderness, hemihypertrophy, Gallop rhythum and basal crepts.**
Spine, eye & genital exam are normal.

DIFFERENTIALS

Age < 2 yr	Age > 2 yr
Neuroblastoma NHL Rhabdomyosarcoma	Wilm's Tumor Germ cell tumor Neuroblastoma

INVESTIGATIONS

NEUROBLASTOMA	WILM'S TUMOR
Most common abdominal mass Crosses midline Embryonic CA of peripheral sympathetic nervous system	2nd most common abdominal mass Doesn't Cross midline Renal tumor
CBC, PT,PTTK, LFTs, ESR, LDH **VMA, HVA** **Neuron specific enolase** **Imaging** (X-ray Abdomen, USG Abdomen, CT/MRI Abdomen) **BMA** (Small, round blue tumor)	**CBC, Urine RE** **CT Scan abdomen** **Bone Scan** **IVU, Renal angiogram** **HRCT Chest** **MRI Brain**

TREATMENT

I. MDT (Multi-disciplinary team approach)
II. PARENTAL COUNSELING

ABDOMINAL MASS

1) General Look
2) Abdominal exam
3) Relevant GPE

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [**child lying flat without pillow** /parent's lap]
d. Exposure: *'Sir Ideally I would like to expose from Mid chest to mid thigh but keeping in view the dignity of child I will expose as required'* (exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Sick/healthy looking
2) Cannula/NG/Foley's/Oxygen
3) Dysmorphism , Hemihypertrophy (Wilm's tumor)
4) Skin rashes, purpura, petechiae, scratch marks
5) Respiratory rate (from foot end)

STEP II: ABDOMINAL EXAM

See Abdominal system exam for details (examine mass in detail: *see Lump/swelling exam*)

STEP III: RELEVANT GPE

Hands	Clubbing, nail changes, Pallor
Pulse	Rate, rhythm, volume, character
BP	V.V.Important ***
BCG scar	Presence
Head & Neck	Hair Eyes [Pallor, Aniridia (Wilm's tumor), Raccoon eyes + opsomyoclonus (Neuroblastoma)] Mouth (oral hygiene) (Tongue)(Angular stomatitis, glossitis or an oral ulcer)
Lymph nodes	Cervical, axillary and inguinal lymph nodes
CVS	Auscultate heart for murmur/failure
Chest	Basal crepts, sternal tenderness
Leg	Pedal edema
Back	Lung bases, Sacral edema, **BMA mark**
Genitalia	*'Sir, I would like to examine genitalia for tanner staging or scrotal swelling'*

Redress the child and say thank you!

3) Test other family members (Hemophilia/carriers)

PROGNOSIS & COUNSELLING

Same as supportive care

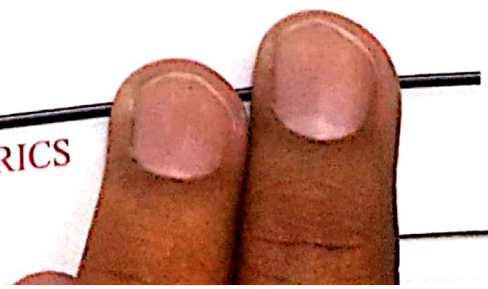
Long-term complications

1) Chronic arthropathy
2) Inhibitor formation
3) Transfusion-transmitted infectious diseases

TIPS & TRICKS

1) Usually the command is GPE or Locomotor system examination (Prioritize according to command).
2) Exposure is very important.

NOTES



assessment before discontinuation of therapy	20 IU/kg every other day for a total of 10-14 days	asymptomatic 40-50 IU/kg every other day for a total of 10-14 days
Major surgery, lifethreatening hemorrhage	**50-75 IU/kg** factor VIII **25 IU/kg q8-12h** (Target trough level >50 IU/dL) for 5-7 days **50 IU/kg q24h** (Target trough level >25 IU/dL) for 7 days	**120 IU/kg** factor IX **50-60 IU/kg** every 12-24 hr (Target >40 IU/dL) for 5-7 days, then (Target >30 IU/dL) for 7 days
Minor	20 IU/kg factor VIII	40 IU/kg factor IX concentrate
Mouth, deciduous tooth, or tooth extraction	20 IU/kg factor VIII concentrate; Antifibrinolytic therapy; Remove loose deciduous tooth	40 IU/kg factor IX concentrate‡; Antifibrinolytic therapy§; Remove loose deciduous tooth
Epistaxis	Apply pressure for 15-20 min; Pack with petrolatum gauze; Antifibrinolytic therapy; 20 IU/kg factor VIII (Persistent)	Apply pressure for 15-20 min; Pack with petrolatum gauze; Antifibrinolytic therapy; 30 IU/kg factor IX (Persistent)
Hematuria	Bed rest; 1.5× maintenance fluids; Persisting >2days 20 IU/kg factor VIII concentrate; ± Prednisone (unless patient is HIV-infected)	Bed rest; 1.5× maintenance fluids; Persisting >2days 40 IU/kg factor IX concentrate ± Prednisone (unless patient is HIV-infected)
Prophylaxis	20-40 IU/kg factor VIII every other day to achieve a trough level ≥1%	30-50 IU/kg factor IX every 2-3 days to achieve a trough level ≥1%

†For mild or moderate hemophilia A : Desmopressin 0.3 µg/kg *(releases factor VIII from body)* Concentrated intranasal form : 150 µg (1 puff) for <50 kg
and 300 µg (2 puffs) for >50 kg

‡Stated doses = Recombinant factor IX; for plasma-derived = Use 70% of the stated dose.

§Do not give antifibrinolytic therapy until 4-6 hr after a dose of prothrombin complex concentrate.

††For repeated doses of factor IX = use highly purified, specific factor IX concentrate.

Prophylaxis (Recommended by The National Hemophilia Foundation)

Initiated with the first joint hemorrhage

Insertion of a central venous catheter in young children

Treatment provided every 2-3 days to maintain a trough level ≥1% before the next infusion

Secondary prophylaxis for target joints in older child Not given primary prophylaxis

Expensive but highly effective in preventing /greatly limiting the degree of joint pathology;

Complications

1) Central line infection
2) Thrombosis

V. FOLLOW UP/MONITORING OF DISEASE ACTIVITY

At Comprehensive hemophilia care centers

(Team of physicians, nurses, orthopedists, physical therapists, and psychosocial workers)

1) Patients exposed to plasma-derived products : Screen for hepatitis B and C, HIV, LFTs
2) Screening for inhibitors

		6) Aminocaproic acid/Tranexamic acid		

b) Details

I. MDT (Multi-disciplinary team approach)

Pediatrician	Clinical Hematologist	Psychologist	Orthopedic surgeon
Physical therapist	Dietician	Social worker	Psychiatrist

II. SUPPORTIVE TREATMENT

1) Avoid trauma
2) Avoiding high-risk behaviors
3) Avoid violent contact sports
4) Avoid aspirin and other NSAIDs affecting platelet function
5) Use of car seats, seatbelts, and bike helmets
6) Early psychosocial intervention (Balance between overprotection and permissiveness)
7) Vaccinations against hepatitis B
8) Patients exposed to plasma-derived products : Screen for hepatitis B and C, HIV, LFTs

2. Specific therapy

1) Factor VIII/IX concentrate
2) Cryoprecipitate (factor VIII deficiency)
3) FFP/ Cryosupernatant (factor IX deficiency)
4) Whole fresh blood

} If Factor VIII/IX Not available

5) Desmopressin acetate
6) Aminocaproic acid/Tranexamic acid

	Target
Mild to moderate bleeding	35-50% activity
Life threatening / major hemorrhages	100% activity

Calculation of the dose of recombinant factors

Dose of rFVIII IU = 0.5 x body weight(kg) x % desired rise in rFVIII activity

Dose of rFIX IU = 1.4 x body weight(kg) x % desired rise in FIX activity

TYPE OF HEMORRHAGE	HEMOPHILIA A	HEMOPHILIA B
Major	50 IU/kg stat then 25 IU/kg	100 IU/kg stat then 50 IU/kg
Hemarthrosis (Orthopedic aspiration if hip involved…risk of AVN)	Day 1: **50-60 IU/kg** FVIII Day 2, 3, 5: **20-30 IU/kg** until joint function is normal + Alternate day treatment (7-10 day) Consider prophylaxis.	Day 1: **80-100 IU/kg** Day 2,4 : **40 IU/kg** + Alternate day treatment (7-10 days) Consider prophylaxis
Muscle /large subcutaneous hematoma	50 IU/kg factor VIII concentrate; 20 IU/kg alternate day till resolved	80 IU/kg factor IX concentrate‡: every 2-3 days till resolved
Iliopsoas hemorrhage *Repeat radiologic*	50 IU/kg factor VIII concentrate, 25 IU/kg every 12 hr until asymptomatic	120 IU/kg factor IX concentrate 50-60 IU/kg every 12-24 hr to (Target >40 IU/dL) till patient is

INVESTIGATIONS

a) Tabulated Overview

I. FOR SCREENING	II. DIAGNOSTIC	III. TO RULE OUT COMPLICATIONS
1. **APTT** 2. **Clotting time** 3. **Mixing of plasma** 4. **Quantitative Bethesda assay** 5. ProThrombin time 6. Thrombin time 7. Platelet count 8. Bleeding time 9. vWF activity	1. Specific factor assay 2. Molecular techniques (Antenatal) 3. Ratio of factor VIII to von Willebrand factor	1. Abdominal radiograph 2. Ultrasonography Abdomen 3. CT scan Abdomen

b) Details

I. FOR SCREENING

1. **PTT**(Raised 2-3 times in severe hemophilia)
2. **Clotting time (Prolonged)**
3. **Mixing of normal plasma with patient plasma** : correction of PTT value (Reverse if inhibitors to factor VIII or IX are present)
4. **Quantitative Bethesda assay for inhibitors**
   Antibodies against active clotting site (25-35% of pt receiving infusions of factor VIII or IX)
5. ProThrombin time (N)
6. Thrombin time (N)
7. Platelet count (N)
8. Bleeding time (N)
9. vWF activity (N)

II. DIAGNOSTIC

1. **Specific factor assay** for VIII and IX
2. **Molecular techniques** via amniocentesis (for mutation in factor VIII gene) (Antenatally)
3. **Ratio of factor VIII to von Willebrand factor** to diagnose carriers

III. TO RULE OUT COMPLICATIONS

1. **Abdominal radiograph** (Psoas sign ipsilaterally & contra-lateral shifted colon gas: *iliopsoas bleed*)
2. **Ultrasonography Abdomen**
3. **CT scan Abdomen** (Hematoma in muscle & anterior translocation of kidney : *iliopsoas bleed*)

TREATMENT

Early, appropriate therapy is the hallmark of excellent hemophilia care.

a) Tabulated Overview

MDT	Supportive R_x	Specific R_x	R_x of Complications	F/UP
	1) Lifestyle changes 2) Psychosocial 3) Vaccines 4) Screening	1) Factor VIII/IX concentrate 2) Cryoprecipitate 3) FFP/Cryosupernatant 4) Whole fresh blood 5) Desmopressin acetate	1) Ch Arthropathy 2) Inhibitor formation 3) Transfusion related infections	

STEP V: RELEVANT SYSTEMIC EXAM

1) **Abdomen** (Hepatosplenomegaly in hepatic disease, tenderness in retroperitoneal bleed)
2) **Chest** (Basal crepts)
3) **CVS** (Gallop rhythum, pericardial rub, murmur of RHD)

Redress the child and say thank you!

OFFER (DO IF TIME PERMITS/LONG CASE)

1) Vitals
2) Anthrpometric measurements
3) Fundoscopy (for hemorrhage)
4) Neurological exam (For focal deficit in ICB)

DESCRIPTION

General look	I have examined _____(name) __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He has **pallor** but no **abdominal distension, distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with Respiratory Rate = ___ /min, BP ____mmHg Pulse is ___/min Regular in rhythm and normal in volume & character
Anthropometry	Weighing ____kg with Height of ____cm & Fronto-Occipital Circumference of _____cm. (I would like to plot them on centile charts)
Gait	**Gait & Back** examinations are normal.
Local exam of swelling	A **diffuse** swelling is noted at **Right knee/elbow** **Overlying skin** is of normal color, temperature & texture with/without discharge/sinus. Swelling is **non-tender/ tender** with **rounded margins, smooth / irregular surface & soft / firm / hard consistency.** It is **non-mobile / mobile & attached** to Skin/Underlying Structures No **pulsations / thrill or bruit** is noted. **Rest of the joint examination** is unremarkable.
GPE	There is no evidence of **clubbing, nail changes, thumb or radial anomalies, rash, petechiae, bruise, jaundice, hepatosplenomegaly, abdominal tenderness, lymphadenopathy, Gallop rhythum, basal crepts & hyperpigmentation.** **BCG** scar mark is present with normal **oral cavity** and **dentition**

DIFFERENTIALS

For short case	For Long case
1) **Hemophilia** 2) **Septic arthritis** 3) **Trauma** 4) **Tuberculosis of joint** 5) **JIA**	1) **Hemophilia** 2) **Severe ITP** 3) **Platelet function disorders** 4) **Type 3 vWD** 5) **Vit K deficiency**

HEMOPHILIA

1) General Look
2) Gait ,Back & Anthropometry
3) GPE
4) Locomotor exam
5) Relevant systemic exam

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building (Talk to child to assess IQ)**
c. **Position patient [child lying flat without pillow** /parent's lap]
d. **Exposure: EXPOSE, EXPOSE, EXPOSE** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Sick/healthy looking
2) Cannula/NG/Foley's/Oxygen
3) Pallor
4) Aides (sticks/wheel chair)
5) Respiratory rate (from foot end)

STEP II: GAIT, BACK & ANTHROPOMETRY

Ask: 'Bachaa Chal saktaa hai?' (Proceed if Yes: Otherwise go to Step III)
Rapid motor scan & Gait examination
Anthropometry *(Do if command is GPE: otherwise skip)*

STEP III: GPE

Hands	Clubbing, nail changes, thumb or radial anomalies, Pallor
Pulse	Rate, rhythm, volume, character
BCG scar	Presence
Head & Neck	Hair Eyes (pallor, jaundice, conjunctival hemorrhage) Mouth (oral hygiene) (Tongue)(Angular stomatitis, glossitis or an oral ulcer) JVP
Lymph nodes	Cervical, axillary and inguinal lymph nodes
Leg	Pedal edema
Back	Lung bases, Sacral edema, **BMA mark**
Genitalia	*'Sir, I would like to examine genitalia for tanner staging or scrotal swelling'*

STEP IV: LOCOMOTOR SYSTEM

See Locomotor system exam for details

II. FOR DIAGNOSIS

1) **Platelet count :** Severe thrombocytopenia (platelet count <20 × 10⁹/L) in ITP
2) **Platelet morphology :** Platelet size is normal or increased (↑platelet turnover) in ITP
3) **Platelet antibody testing (IgG, IgM)** for ITP
4) **VWF antigen** (VWF : Ag) to measures the total amount of VWF protein
5) **Bone marrow aspirate and trephine:** Aplasia (marrow cellularity <25%), ITP (increased numbers of megakaryocytes)
6) **Cytogenetics and chromosomal breakage studies :** To detect Fanconi's anemia or dyskeratosis congenital

III. TO RULE OUT COMPLICATIONS

CT Scan Brain (for ICB)

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELING

Depends on cause

III. TREATMENT OF ACUTE PROBLEMS

Strict Anti-septic measures
Depending on severity: **RBC +/– platelet transfusion**
Treat Infections with **antibiotics, antifungal**

IV. SPECIFIC TREATMENT

ITP	**Tranexamic acid 20–25mg/kg tds for <5 days** (Moderate bleed) **IVIG @ 0.8-1 g/kg/day for 1-2 days** **Prednisone of 1-4 mg/kg/24 hr** **IV anti-D therapy** For Rh-positive **50-75 µg/kg (Rhogam)** **Rituximab** **Splenectomy**
Von willebrand disease	**Antifibrinolytics** (For mucosal bleeding, all types of VWD) **Cryoprecipitate** **Desmopressin** (Type 1) **von Willebrand factor concentrates** (type 3,2, severe 1)
Aplastic anemia	Remove or **treat underlying cause**, e.g. drugs/infections **Hematopoietic stem cell transplantation** (HSCT) from a sibling with identical HLA and compatible mixed lymphocytes.(survival rate >80%) **Immunosuppression**, e.g. rabbit anti-thymocyte globulin followed by ciclosporin is best second line therapy for those with no BMT donor **Corticosteroids and androgens** (oxymetholone) in Fanconi anemia

TIPS & TRICKS

...t hidden places.

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals
2) Anthrpometric measurements
3) Fundoscopy (for hemorrhage)

DESCRIPTION

General look	I have examined _____(name) __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He has **pallor** but no **abdominal distension, distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with Respiratory Rate = ___ /min, BP ____mmHg Pulse is ___/min Regular in rhythm and normal in volume & character
Inspection	Abdomen is of **normal shape** with **central umbilicus.** It is **moving with respiration** There are no **prominent veins, scars, striae, rash, petechiae** or **scratch marks.**
Palpation	Abdomen is **soft, non-tender** without evidence of **visceromegaly** and **lymphadenopathy.**
Percussion	Bladder is not percussable. **Shifting dullness** is absent.
Auscultation	**Bowel sounds** are audible. There is **no bruit.**
Genitalia	**Genetalia** are pre pubertal. **Inguinal nodes** are not palpable & **Hernial orifices** are intact.
Back, leg	There is no evidence of **basal crepts, sacral** or **pedal edema.**
GPE	There is no evidence of **clubbing, nail changes, thumb or radial anomalies, petechiae, bruise, bleed, joint swelling, lymphadenopathy, jaundice & hyperpigmentation.**
	BCG scar mark is present with normal **oral cavity** and **dentition**
	CVS, Chest, Gait & Joint examinations are unremarkable

DIFFERENTIALS

1) **ITP**
2) **Von-willebrand disease**
3) **Aplastic anemia/Fanconi anemia** (if features are suggestive)
4) **Pre-leukemic leukemia**

INVESTIGATIONS

I. SUPPORTIVE

1) **Hemoglobin value, WBC count, and differential count**
2) **PT, PTTK** to rule out other causes of coagulopathy (N)
3) **BT** (prolonged in ITP)
4) **Direct antiglobulin test (Coombs)** if there is unexplained anemia to rule out Evans syndrome (autoimmune hemolytic anemia and thrombocytopenia) or before instituting therapy with IV anti-D
5) **Blood grouping** (to assess Rh positivity)

APLASTIC/FANCONI ANEMIA/ PETECHIAE / BRUISES

1) General Look
2) Abdominal exam
3) Relevant GPE
4) Gait & Joint examination

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building (Talk to child to assess IQ)**
c. **Position patient [child lying flat without pillow /parent's lap]**
d. **Exposure: EXPOSE, EXPOSE, EXPOSE** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Sick/healthy looking
2) Cannula/NG/Foley's/Oxygen
3) Pallor, Hyperpigementation (Café-au-lait spots)
4) Skin rashes, purpura, petechiae, Bruise (Distribution, size, colour)
5) Respiratory rate (from foot end)

STEP II: ABDOMINAL EXAM

See Abdominal system exam for details (must include palpation of kidneys)

STEP III: RELEVANT GPE

Hands	Clubbing, nail changes, thumb or radial anomalies, Pallor
Pulse	Rate, rhythm, volume, character
BCG scar	Presence
Head & Neck	Hair Eyes (pallor, jaundice, conjunctival hemorrhage) Mouth (oral hygiene) (Tongue)(Angular stomatitis, glossitis or an oral ulcer) JVP Thyroid
Lymph nodes	Cervical, axillary and inguinal lymph nodes
CVS	Auscultate heart for murmur/failure
Leg	Pedal edema
Back	Lung bases, Sacral edema, **BMA mark**
Genitalia	*'Sir, I would like to examine genitalia for tanner staging or scrotal swelling'*

STEP IV: GAIT & JOINT EXAMINATION

1) Gait & rapid motor scan to rule out Intra-cranial bleed
2) Joint examination for Hemarthrosis

5. Transfusion therapy 6. Iron overload monitoring 7. Chelation therapy 8. Hydroxyurea 9. Splenectomy 10. HSCT	pre-natal diagnosis		

TIPS & TRICKS

1) Don't miss all lymph node stations.

NOTES

	Kala-azar IE	T.B. (icterus is due to drugs)		HIV	Dengue IE
2.Hemato-logical	Thalassemia Sickle cell HS	Thalassemia Sickle cell HS	Thalassemia	Thalassemia	Thalassemia HSP Ch. ITP
3. Malignancy	Leukemia Lymphoma	Leukemia Lymphoma	Leukemia Lymphoma	Leukemia Lymphoma	Leukemia Lymphoma
4. Metabolic	Wilson Gaucher	Galactosemia Alpha-1 antitrypsin deficiency	Galactosemia Alpha-1 antitrypsin deficiency	-	Gaucher Nieman pick disease
5. Misc	SLE Cirrhosis with portal HTN	Drugs Cirrhosis with portal HTN	Drugs Cirrhosis with portal HTN CCF	SLE JIA	Cirrhosis with hypersplenism

KEY

HSM=Hepatosplenomegaly, **LN**=Lymphadenopathy, **Bleed**=Bleeding manifestation

INVESTIGATIONS	
CBC	For degree of pallor, blood indices, evidence of infection
Peripheral film	For sickle cell, HS
MP slide	Thick and thin film for for malaria
Blood culture	For bacterial infections
Serology	For Leishmaniasis
LFTs	To rule out Jaundice & Hepatic involvement
Hb electrophoresis	For sickle cell disease, Thalassemia
BMA	For Leukemia/Lymphoma

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELING

Prognosis depends on cause

III. TREATMENT OF ACUTE PROBLEMS

RCC transfusion

IV. SPECIFIC TREATMENT

Treat the cause

THALASSEMIA TREATMENT OVERVIEW

Patient Oriented	Family Oriented	R_x of Complications	Monitoring
Non-Pharmacological 1. Nutrition 2. Vitamins & minerals 3. Physical activity 4. Psychosocial support *Pharmacological*	a) Parental counseling b) Genetic counseling c) Prevention : screening &	1.Hep B & C 2. CLD 3. Bone(Osteopenia/ Osteoporosis) 4.Cardiac 5.Endocrine	Monthly 6 monthly Yearly As required

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals
2) Anthrpometric measurements
3) Fundoscopy (for hemorrhage)

DESCRIPTION

General look	I have examined _____(name) __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He has **pallor** and **abdominal distension** but no **distress** or **dysmorphism/Thalassemic facies.**
Vitals	He is afebrile (to touch) with Respiratory Rate = ___ /min, BP ____mmHg Pulse is ___/min Regular in rhythm and normal in volume & character
Inspection	Abdomen is **distended** with **prominent veins** and central **everted umbilicus, moving with respiration** **Direction of venous blood** is away from umbilicus There are no **scar, striae, rash, petechiae** or **scratch marks.**
Palpation	Abdomen is **soft , non-tender** **Liver** is palpable ___cm below RCM, having a **total span** of ___cm with upper **border** percussable in ___ ICS. It is **non-tender,** with **sharp margins,** firm **consistency,** smooth **surface** and **Left lobe** of liver is not palpable. **Spleen** is palpable __ cm below LCM along its axis. It is **non-tender, with regular margins,** firm **consistency,** smooth **surface** and **notch** is (not) palpable. No evidence of other visceromegaly.
Percussion	Bladder is not percussable. **Shifting dullness** is positive.
Auscultation	**Bowel sounds** are audible. There is **no bruit.**
Genitalia	**Genetalia** are pre pubertal. **Inguinal nodes** are not palpable & **Hernial orifices** are intact.
Back, leg	There is no evidence of **basal crepts, sacral** or **pedal edema.**
GPE	There is no evidence of **clubbing, nail changes, thumb or radial anomalies, petechiae, bruise, bleed, joint swelling, lymphadenopathy, jaundice & hyperpigmentation.**
	BCG scar mark is present with normal **oral cavity** and **dentition**
	CVS and **Chest** examinations are unremarkable

DIFFERENTIALS

1) **Chronic infections** (Malaria, Enteric fever, Leishmaniasis, Disseminated TB)
2) **Hemolytic anemias** (e.g. Thalassemia)
3) **Malignanacy** (ALL/NHL)

AJ'S SUPER TABLE OF HSM DIFFERENTIALS

Groups	HSM+Pallor	HSM+Jaundice	HSM+Ascites	HSM+LN	HSM+Bleed
1.Infections	Malaria Disseminated T.B.	Hepatitis Malaria Disseminated	Hepatitis Abdominal T.B.	Disseminated T.B. Kala-azar	Septicemia TORCH infections

ANEMIA WITH HEPATOSPLENOMEGALY

1) General Look
2) Abdominal exam
3) Relevant GPE

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [**child lying flat without pillow** /parent's lap]
d. Exposure: *'Sir Ideally I would like to expose from Mid chest to mid thigh but keeping in view the dignity of child I will expose as required'*(exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Sick/healthy looking
2) Cannula/NG/Foley's/Oxygen
3) Thalassemic facies, Jaundice, pallor
4) Skin rashes, purpura, petechiae, scratch marks
5) Respiratory rate (from foot end)

STEP II: ABDOMINAL EXAM

See Abdominal system exam for details (must include inguinal lymph nodes)

STEP III: RELEVANT GPE

Hands	Clubbing, nail changes, thumb or radial anomalies, Pallor
Pulse	Rate, rhythm, volume, character
BCG scar	Presence
Head & Neck	Hair Eyes (pallor, jaundice) Mouth (oral hygiene) (Tongue)(Angular stomatitis, glossitis or an oral ulcer) JVP Thyroid
Lymph nodes	Cervical, axillary and inguinal lymph nodes
CVS	Auscultate heart for murmur/failure
Leg	Pedal edema
Back	Lung bases, Sacral edema, **BMA mark**
Genitalia	*'Sir, I would like to examine genitalia for tanner staging or scrotal swelling'*

Redress the child and say thank you!

<table>
<tr><td></td><td>it can be associated with significant complications.</td></tr>
<tr><td>End-stage liver disease (liver failure)</td><td>Transplantation
1) Success rate >85%
2) Prolongs life
3) Might correct metabolic error in diseases such as α1-anti-trypsin deficiency, tyrosinemia, and Wilson disease.
4) Scarcity of donors of small livers severely limits application of liver transplantation for infants and children. Use of reduced-size transplants & living donors increases ability to treat small children successfully.</td></tr>
<tr><td>Follow-up</td><td>Dietary counseling, anthropometry and monitoring of serum and urinary electrolyte concentrations</td></tr>
<tr><td>Prognosis</td><td>
<table>
<tr><th>Sporadic idiopathic neonatal hepatitis</th><th>Familial idiopathic neonatal hepatitis</th></tr>
<tr><td>60-70% recover</td><td>20-30% recover</td></tr>
<tr><td>5-10% have persistent fibrosis or inflammation</td><td></td></tr>
<tr><td>Smaller percentage have severe liver disease/cirrhosis</td><td>10-15% acquire CLD with cirrhosis</td></tr>
<tr><td>Infants usually die early due to hemorrhage or sepsis</td><td>May need liver transplantation</td></tr>
</table>
</td></tr>
</table>

TIPS & TRICKS

1) Be opportunistic in examining infant. If infant appears irritable, auscultate first.

NOTES

	alternative diagnosis

TREATMENT

MDT: Pediatrician, pediatric gastroenterologist, pediatric surgeon, dietician

CLINICAL IMPAIRMENT	MANAGEMENT
Malnutrition : malabsorption of dietary long-chain triglycerides	Replace with dietary formula or supplements containing medium chain triglycerides
Fat-soluble vitamin malabsorption	Vit K : 2.5-5.0 mg every other day as water-soluble derivative of menadione
	Vit E: Replace with 50-400 IU/day as oral α-tocopherol or TPGS (D-tocopherol polyethylene glycol 1,000 succinate)
	Vit A : 10,000-15,000 IU/day as Aquasol A
	Vit D: Replace with 5,000-8,000 IU/day of D2 or 3-5 µg/kg/day of 25-hydroxycholecalciferol
Deficiency of water-soluble vitamins	Supplement with twice the recommended daily allowance
Micronutrient deficiency	Calcium, phosphate, or zinc supplementation
Retention of biliary constituents e.g. cholesterol (itch /xanthomas)	Choleretic bile acids (ursodeoxycholic acid, 15-30 mg/kg/day) : can increase bile flow or interrupt the enterohepatic circulation of bile acids, can also lower serum cholesterol
Pruritus refractory to medical therapy	**Partial external biliary diversion** **Technique**: resecting a segment of intestine to be used as a biliary conduit. One end of the conduit is attached to the gallbladder and the other end is brought out to the skin, forming a stoma. **Drawback**: need to use an ostomy bag
Ascites	1) Rule out spontaneous bacterial peritonitis 2) Restrict sodium intake to 0.5 g (1-2 mEq/kg/24 hr) 3) Restrict fluid if renal output is inadequate 4) Diuretics (if above is ineffective) Spironolactone (diuretic of choice) *(Aldactone 100mg)* (3-5 mg/kg/24 hr in 4 doses) (1mg/kg/dose QID) Spironolactone + furosemide *(Tab Spiromide 20mg)* Spironolactone + thiazide Patients with ascites but without peripheral edema are at risk for reduced plasma volume and decreased urine output during diuretic therapy. 5) Paracentesis and intravenous albumin infusion
Portal hypertension (variceal hemorrhage & Hypersplenism)	Ascertain cause of bleeding (Variceal/gastritis/APD) via endoscopy Blood transfusion (If volume depleted) Sclerotherapy/endoscopic variceal ligation may be useful palliative measures & superior to surgical alternatives. *Balloon tamponade is not recommended in children because*

3) Fundoscopy (for cherry red spots in GSD, cataract, chorioretinitis)
4) Developmental assessment
5) Neurological exam

DESCRIPTION

See description of Chronic Liver disease (Elder child)

DIFFERENTIALS

1) **Biliary atresia**
2) **Choledochal cyst**
3) **Neonatal hepatitis**
4) **PFIC**
5) **Glycogen storage disorder**
6) **TORCH infection**
7) **Tyrosinemia**

INVESTIGATIONS

TEST	RATIONALE
1) Confirming cholestasis	
Assessment of stool color (does the baby have pigmented or acholic stools?)	Indicates bile flow into intestine
Serum bilirubin fractionation (esp.conjugated)	Indicates cholestasis
ALP, Gamma GT, 5' Nucleotidase	Indicate cholestasis
Urine and serum bile acids measurement	Confirms cholestasis; might indicate inborn error of bile acid biosynthesis
2) Assessing severity of liver disease	
Hepatic synthetic function (albumin, coagulation profile)	Indicates severity of hepatic dysfunction
3) Finding etiology& type of cholestasis	
a)Infections	
CBC, CRP	To rule out sepsis
Viral serology (echo , herpes, A,B,C)	Viral hepatitis
TORCH screen (CMV), CSF VDRL test	Congenital infections (TORCH, Syphilis)
b) Genetic causes	
α1-Antitrypsin phenotype	Suggests/excludes PiZZ (protease inhibitor)
Sweat chloride and mutation analysis	Suggests (or excludes) cystic fibrosis
c) Meatbolic	
Thyroxine and TSH	Suggests (or excludes) endocrinopathy
Urine and serum amino acids	Suggests (or excludes) metabolic liver disease
Urine reducing substances	Suggests (or excludes) Galactosemia
d) Surgical causes	
Ultrasonography of bile ducts and gallbladder	Suggests (or excludes) choledochal cyst; Triangular cord sign (biliary atresia)
Hepatobiliary scintigraphy	Documents bile duct patency or obstruction
Liver biopsy	Distinguishes biliary atresia; suggests

CHRONIC LIVER DISEASE (INFANT)

1) General Look
2) Abdominal exam
3) Relevant GPE

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [**child lying flat without pillow /Parent's lap**]
d. Exposure: (Shirt off, trousers rolled up) (Seek parent's help to undress) Beware of hypothermia
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Sick/healthy looking
2) Cannula/NG/Foley's/Oxygen
3) Dysmorphic features (Alagille syndrome), Jaundice
4) Skin rashes, purpura, petechiae, scratch marks

STEP II: ABDOMINAL EXAM

See Abdominal system exam for details

STEP III: RELEVANT GPE

Hands	Leuconychia, Clubbing, Palmar erythema, Pallor, Wrist widening
Pulse	Rate, rhythm, volume, character
BCG scar	Presence
Head & Neck	Hair; Anterior Fontanelle (for <2 yr), Microcephaly (TORCH) Eyes (icterus, corneal ulcer, pallor, Bitot spots, Cataract) Mouth (oral hygiene) (Tongue)(Angular stomatitis, glossitis or an oral ulcer) Thyroid
Lymph nodes	Cervical, axillary and inguinal lymph nodes
CVS	Auscultate heart for murmur (TORCH, Alagille syndrome)
Chest	Rachitic rosary
Leg	Pedal edema
Back	Lung bases, Sacral edema, Wasted buttocks
Nappy	Color of stool

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals
2) Anthrpometric measurements + MUAC

RECENT ADVANCES

i. Gene therapy
ii. Receptor-based targeted enzyme replacement therapy

TIPS & TRICKS

1) Use dipping method to palpate tense and distended abdomen in ascites.

NOTES

Broad spectrum antibiotics (cefotaxime, Tazobactum)

7. Salt and water retention/ascites (poor prognostic sign)
Sodium restriction (less than 2 mmol/kg/day) and salt restriction to 0.3–0.5 g/day
(Loss of 100–150 mEq of salt = loss of 1 litre of water) *Aim = 250 g weight loss/day*

Spironolactone(aldosterone antagonist) (K sparing diuretic)
Salt-poor albumin and frusemide
Abdominal tap (may be diagnostic but is otherwise of little value)

Haemodialysis or haemofiltration (ARF/hepatorenal failure)

8. Pruritus (esp in biliary hypoplasia)
Ursodeoxycholic acid (Treatment of choice)
Add on: Rifampicin,cholestyramine, phenobarbitone, ondansetron and naltrexone

9. Drugs
Avoid ATT, phenytoin, sulphonamides, erythromycin and paracetamol

10. Liver transplantation
Indications:
1. Failure of hepatic synthetic function
2. Poor quality of life (e.g. intractable pruritis, lethargy, anorexia, recurrent infections),
3. Intractable malnutrition /FTT
4. Refractory ascending cholangitis
5. Hyperammonaemia(IEMs)
6. Encephalopathy
7. Oesophageal varices from portal hypertension, and hypersplenism.
8. Specific diseases:
   i. Extrahepatic biliary atresia (EHBA): over 50% of LTx.
   ii. IEMs (10–15% of LTx) include AT deficiency, CF, galactosaemia, GSDs type IA and IV, mitochondrial functional defects (e.g. defects of fatty acid beta-oxidation), some organic acidaemias, tyrosinaemia, urea cycle defects and WD.
   iii. Acute hepatic necrosis (around 10%)
   iv. Cirrhosis from chronic active hepatitis or primary biliary cirrhosis (under 10%)
   v. Cholestatic liver disease, other than EHBA (under 5%)
   vi. Primary hepatic malignancy (2%)

Absolute Contraindication
Irreversible extrahepatic disease (e.g.HIV, irreversible brain damage, incurable malignancy)

11.Specific Therapy
   i. Treatment of Hep B & C (Interferon-A +/– Ribavirin)
   ii. Autoimmune hepatitis (Prednisolone +/– Azathioprine)
   iii. Replacement: Oral administration of primary bile acids
   iv. Chelation and antioxidant mixtures (Neonatal iron storage disease)
   v. Metabolic inhibitors (NTBC) in tyrosinaemia
   vi. Enzymes inducer(Phenobarbitone) in Crigler–Najjar syndrome type II
   vii. Dietary restriction of galactose (galactosaemia)
   viii. Molecular manipulation (inhibition of polymerisation of alpha-1-antitrypsin)

LTx	10. Liver transplant
Others	11. Disease specific

MDT

Pediatrician	Gastroenterologist	Ophthalmologist	Surgeon
Physical therapist	Dietician	Social worker	Nurse clinician
Psychologist	Psychiatrist		

1.Supportive

(ALP – OF - TTNM Care)

Monitor: Weight, serum albumin, PT, serum bilirubin (total and conjugated),

Psychosocial support is important

2. Encephalopathy

(Aim= decreasing nitrogenous load to the bowel + bacterial production of ammonia)

Review precipitating factors (infection, large GI bleed, excess protein intake, electrolyte imbalance & diuretics)

Restrict protein(2 g/kg/day)

Substitution with branch-chained amino acids (leucine, isoleucine and valine)

Oral lactulose & neomycin + Tab Rifaxamine (Rifaxa) 550mg + soluble fibre pectin

3. Nutrition

High energy (120–150% of the recommended daily amount)

Moderately high protein intake(3–4 g/kg/day)

Branched chain amino acid enriched formulations (do not require the liver for metabolism)

Enteral feeding (if anorectic)/ Nocturnal nasogastric enteric feeding

TPN (If enteral feeding is not tolerated)

Fat-soluble vitamins(A, D, E, K) RDA X 3times

Zinc, iron and calcium

4. Portal hypertension, varices and varicealhaemorrhage (evaluated endoscopically)

Intensive-care management

IV fluids and blood products

Vit K , Platelets, FFP

Intravenous Vasopressin/Octreotide/Glypressin to reduce portal pressure

Band ligation (Success 70–100%, rebleeding 15–30%) or **Sclerotherapy**(once stable)

Balloon tamponade: modified Sengstaken–Blakemore tube + IV vasopressin for 24–48hr(Complications: Pulmonary aspiration, oesophageal rupture and suffocation)

Transjugular intrahepatic portosystemic stent shunt (TIPSS) (Success 80–100%)

(Complications = Occlusion of stent, the development of encephalopathy and infection.)

Avoid splenectomy if possible (Risk of infection, increased bleeding from removal of good collateral vessels from the splenic capsule (azygos system) that bypass the lower oesophageal junction vessels.

Prophylactically : Tab Carvedalol

5. Coagulopathy

Vitamin K (2–10 mg/day)

Fresh frozen plasma, cryoprecipitate and platelets (During bleeding)

6. Sepsis

Common in CLD (especially ascending cholangitis and bacterial peritonitis)

Can precipitate encephalopathy, or acute or chronic liver failure

4) Viral hepatitis (elder children only)

INVESTIGATIONS

Screening for liver disease& its nature	**Tests for hepatocellular damage** (AST, ALT) **Tests for biliary excretion** (total and direct bilirubin) **Tests for cholestasis** (GGT, AP,5' nucleotidase) **Tests for liver synthetic function** (Albumin, PT, INR, globulin levels)
Severity?	**ABGs** (for acidosis) **Ammonia** (hyperammonemia/test for detoxification functions) **Albumin levels (serial)**(Marked hypoalbuminemia) **BSR** (for hypoglycemia) **Bilirubin levels (serial)**(Continued hyperbilirubinemia) **CBC**(for thrombocytopenia, leucopenia in hypersplenism) **Electrolytes** (for imbalance e.g. hypo/hypernatremia, hyperkalemia) **Prolonged PT or INR** (unresponsive to parenteral vitamin K)
To ascertain Etiology?	
1. Wilson's disease (WD)	S. cerruloplasmin (low <20mg/dl) Coomb negative hemolytic anemia S. copper levels KF rings (slit lamp) 24 hr urinary copper (>100 microgram/24 hrs) D-penicillamine challenge (>1000 microgram/24 hrs) Quantification of liver copper by liver biopsy (>250 microgram/g dry weight of liver) (Gold standard)
2. Autoimmune chronic hepatitis	High γ-globulin levels Type 1: ASMA (smooth muscle), ANA Type 2: Anti-LKM, Anti-liver cytosolic antibodies Type 3: Anti SLA (soluble liver antibodies)
3. Drugs	History, Drug levels
4. Infections	Hep B , C, CMV serology
5. Storage	Liver Biopsy

TREATMENT

TREATMENT OUTLINE

MDT	
Head	1. Supportive + Psychotherapy 2. Encephalopathy
Oral cavity	3. Nutrition
Esophagus	4. Variceal bleed, Portal HTN
Blood	5. Coagulopathy 6. Sepsis
Abdomen	7. Ascites
Skin	8. Pruritis
Avoid	9. Drugs, diet

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals
2) Anthrpometric measurements + MUAC
3) Fundoscopy
4) Slit lamp examination (KF rings)
5) Developmental assessment
6) Neurological exam
7) Joint exam (for autoimmune hepatitis)

DESCRIPTION

General look	I have examined _____(name) __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He has **clubbing, jaundice** and **abdominal distension** but no **distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with Respiratory Rate = ___ /min, BP ____mmHg Pulse is ___/min Regular in rhythm and normal in volume & character
Inspection	Abdomen is **distended** with **prominent veins** and central **everted/slit like umbilicus, moving with respiration** **Direction of venous blood** is away from umbilicus There are no **scar, striae, rash, petechiae** or **scratch marks**
Palpation	Abdomen is **soft , non-tender** **Liver** is palpable ___cm below RCM, having a **total span** of ___cm with upper **border** percussable in ___ ICS. It is **non-tender,** with **sharp margins,** firm **consistency,** smooth **surface** and **Left lobe** of liver is not palpable. **Spleen** is palpable __ cm below LCM along its axis. It is **non-tender, with regular margins,** firm **consistency,** smooth **surface** and **notch** is (not) palpable. No evidence of other visceromegaly.
Percussion	Bladder is not percussable. **Shifting dullness** is positive.
Auscultation	**Bowel sounds** are audible. There is **no bruit.**
Genitalia	**Genetalia** are pre pubertal. **Inguinal nodes** are not palpable & **Hernial orifices** are intact.
Back, leg	There is no evidence of **sacral** or **pedal edema.**
GPE	There is no evidence of **leuconychia, palmar erythema, wrist widening, joint swelling, lymphadenopathy, petechiae, bruise, spider nevi, pallor** or **signs of micronutrient deficiency.**
	BCG scar mark is present with normal **oral cavity** and **dentition**
	CVS and **Chest** examinations are unremarkable

DIFFERENTIALS

1) Wilson's disease
2) Autoimmune hepatitis
3) Drug induced hepatitis

CHRONIC LIVER DISEASE (ELDER CHILD)

1) General Look
2) Abdominal exam
3) Relevant GPE

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [**child lying flat without pillow** /parent's lap]
d. Exposure: *'Sir Ideally I would like to expose from Mid chest to mid thigh but keeping in view the dignity of child I will expose as required'*(exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Sick/healthy looking
2) Cannula/NG/Foley's/Oxygen
3) Abnormal movements, Jaundice
4) Skin rashes, purpura, petechiae, scratch marks

STEP II: ABDOMINAL EXAM

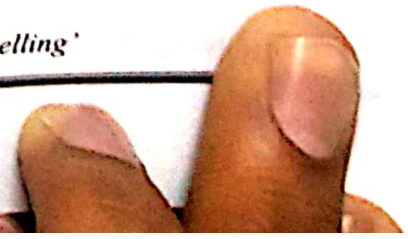
See Abdominal system exam for details

STEP III: RELEVANT GPE

Hands	Leuconychia, Clubbing, Palmar erythema, Astrexis , Pallor, Wrist widening
Pulse	Rate, rhythm, volume, character
BCG scar	Presence
Head & Neck	Hair Eyes (icterus, corneal ulcer, pallor, Bitot spots, Cataract) Mouth (oral hygiene) (Tongue)(Angular stomatitis, glossitis or an oral ulcer) JVP Thyroid
Lymph nodes	Cervical, axillary and inguinal lymph nodes
CVS	Auscultate heart for murmur/failure
Chest	Rachitic rosary
Leg	Pedal edema
Back	Lung bases, Sacral edema, Wasted buttocks
Genitalia	*'Sir, I would like to examine genitalia for tanner staging or scrotal swelling'*

STEP VI: GENITALIA

'Sir, I would like to examine genitalia for tanner staging or scrotal swelling'



3) Anthrpometric measurements + MUAC
4) Stool & urine
5) DRE: Anal fissure/fistulae/skin tags/hemorrhoids/palpable mass

DESCRIPTION

See description of Chronic Liver disease ahead

TIPS & TRICKS

1) Don't check direction of venous blood flow if the veins are not very prominent and dilated.
2) Palpate abdomen sitting at the level of patient with flat hands and looking into eyes of child.
3) Always palpate the surface of liver.
4) If you are confused regarding spleen despite Shorts maneuver, percuss it.
5) To palpate kidneys do not push your hand from the front side, push from the back and palpate from the front hand.
6) If child has massive ascites don't waste your time on shifting dullness, ask the examiner; can you please place your hand on the abdomen and check fluid thrill.
7) If ascites is not massive, check shifting dullness and when you turn the child to recumbent position at the same time check for sacral edema and mark for bone marrow biopsy.
8) While doing superficial and deep palpation; you can have an idea of liver and spleen.

NOTES

3) **Deep palpation** (MovE with Expiration) (soft/tense/guarded & non-tender/tender/ rebound tenderness)
4) **Liver** (Edge, Tenderness, measure, Surface, Consistency, percuss upper margin & measure, Left lobe, Pulsations, Bruit)
5) **Spleen** (Edge, Tenderness, measure, Notch, Surface, Consistency, Pulsations, Bruit) Confirm absence by right lateral tilt & percussion)
6) **Kidneys** (Bilateral: Balloting method)
7) **Bladder**
8) **Lymph nodes**

STEP IV: PERCUSSION

1) **Bladder** (from midline downwards)
2) **Shifting dullness** for ascites
3) **Fluid thrill** (in massive ascites)

STEP V: AUSCULTATION

1) **Bowel sounds** (bowel sounds of normal intensity audible in the right iliac fossa)
2) **Renal bruit** (Left & Right) (Renal artery stenosis)
3) **Hepatic bruit** (Hepatoma)
4) **Venous hum** (epigastric region: turbulent flow of collaterals in portal HTN)
5) **Aortic bruit** (Above & left of umbilicus: aortic narrowing)
6) **Splenic rub** (Perisplenitis)

STEP VI: GENITALIA

'Sir, I would like to examine genitalia for tanner staging or scrotal swelling'

STEP VII: RELEVANT GPE

Hands	Leuconychia, Clubbing, Palmar erythema, Astrexis , Pallor
Pulse	Rate, rhythm, volume, character
BCG scar	Presence
Head & Neck	Hair; Anterior Fontanelle (for <2 yr) Eyes (icterus, corneal ulcer, pallor) Mouth (oral hygiene) (Tongue)(Angular stomatitis, glossitis or an oral ulcer) JVP Thyroid
Lymph nodes	Cervical, axillary and inguinal lymph nodes
CVS	Auscultate heart for failure
Leg	Pedal edema
Back	Lung bases, Sacral edema, Wasted buttocks, mark for bone marrow biopsy

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) B.P.
2) Fundoscopy (for cherry red spots in GSD)

ABDOMINAL EXAM

1) General Look
2) Inspection of Abdomen
3) Palpation
4) Percussion
5) Auscultation
6) Genitalia
7) Relevant GPE

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient [child lying flat without pillow** /parent's lap]
d. **Exposure:** *'Sir Ideally I would like to expose from Mid chest to mid thigh but keeping in view the dignity of child I will expose as required'*(exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Sick/healthy looking
2) Obese/Thin/malnourished
3) Average/tall/short built
4) Cannula/NG/Foley's/Oxygen/Nappies
5) Dysmorphic features, Signs of Dehydration, Jaundice, Pallor, Cyanosis
6) Skin rashes, purpura, petechiae, scratch marks

STEP II: INSPECTION OF ABDOMEN (From foot end & Side)

1) **Respiratory Rate & Pattern** (Abdomino-thoracic/Thoraco-abdominal respiration)
2) **Shape** (full/distended/scaphoid)
3) **Movement** (moving with respiration or not)
4) **Umbilicus** (central & circular/everted/transversely slitted)
5) Prominent **veins**
6) Visible **Pulsations, striae or scars**
7) **Pubic hairs** (absent / of male / female pattern)
8) **Gynaecomastia, spider nevi (CLD)**
9) **Ask to look at side & COUGH:** To look for any hernia

STEP III: PALPATION (sit at the level of patient, warm hands)

'Beta pait main ya taang main kahiin dard tuu nahi hai?' **(Fold knees of patient)**

1) **Venous Direction of flow**
2) **Superficial Palpation** (S shaped) (Look into eyes)

8) CAH
9) Acromegaly

INVESTIGATIONS	
Slit-lamp ophthalmological assessment	Marfan syndrome or homocystinuria
Urine homocystine **Blood homocystine and methionine levels**	Homocystinuria
Electrocardiogram, chest X-ray and Echocardiogram	Marfan syndrome
Plasma somatomedin C concentration and a cerebral CT or MRI scan	Pituitary cause
Skeletal X-rays	McC–A syndrome (bony fibrous dysplasia) or Scoliosis or kyphosis (Marfan,homocystinuria)

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELLING

III.SPECIFIC TREATMENT

Treat the cause

TIPS & TRICKS

1) Use sticky arrows (to be pasted on wall) while taking measurements of the child. Remember to remove it once done.

NOTES

	Tongue • Big (B–W, pituitary gigantism) • Nodular (MEN 2b) **Teeth** • Crowding (homocystinuria) • Separation (pituitary gigantism) **Palate** • High, narrow (Marfan, Sotos, homocystinuria) • Cleft (Marfan)
Chin	• Acne (sexual precocity) • Hair (pubertal staging) • Prognathism (Sotos, B–W, McC–A)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Child's progressive percentile charts (for growth velocity)
3) Parents' heights and onsets of puberty (for MPH & constitutional delay)
4) Visual field examination (for craniopharyngioma)
5) Fundoscopy (for papilledema/ craniopharyngioma)
6) Child's X ray wrist (for Bone age)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
I have examined _____, _____yr old Conscious & cooperative child with **normal/Impaired intellect** and **Marfanoid habitus/ Eunuchoid**
Skeletal features include **Arachnodactyly, Pectus excavatum/carinatum & Scoliosis**
Her/his **total height** is _____cm with **proportionate/disproportionate** Upper & lower segments
Arm span _____, **US:LS** _____
He/she appears to be tall for his/her age and sex but I would like to confirm it by plotting on centile chart. His **weight** is _____kg & **Fronto-occipital circumference** is _____cm
His/her **Vitals** are _____
Manoeuvres for tall stature revealed hypermobility/ normal mobility
Gait is normal. **General physical, Eye, Chest, CVS and abdominal examination** revealed no abnormality/ _____
BCG scar is seen.**SMR** is prepubertal

DIFFERENTIALS

1) Marfan syndrome
2) Homocystinuria
3) Hyperthyroidism
4) Sotos
5) Beckwith–Wiedemann [B–W]
6) Klinefelter
7) Kallman

	Palms • Warm, sweaty (hyperthyroidism) • Pigmented creases (CAH) **Pulse**: collapsing (aortic incompetence in Marfan, hyperthyroidism) **Blood pressure** • Elevated (NF-1, CAH, pituitary gigantism, MEN type 2b with phaeochromocytoma) • Pulse pressure elevated (aortic incompetence with Marfan,hyperthyroidism) **Axillae**: assess pubertal staging for precocity or delay, apocrine secretion, hair, odour
Head	**Size** • Large (Sotos, NF-1) • Small (B–W) **Shape** • Frontal bossing (Sotos) • Prominent occiput (B–W) **Hair** • Receding frontal hairline (Sotos) • Dry, light, sparse (homocystinuria) • Greasy (CAH)
Face	• Asymmetry (B–W, Proteus, NF-1, McC–A) • Fair complexion (homocystinuria) • Malar flush (homocystinuria) • Café-au-lait spots (NF-1, McC–A) • Naevus flammeus (B–W)
Eyes	**Inspect**: • Wearing glasses (myopia with Marfan, homocystinuria) • Blue irides (homocystinuria) • Prominent (B–W, hyperthyroidism) • Exophthalmos (hyperthyroidism) • Hypertelorism (Sotos) • Downslanting (Sotos) • Conjunctival neuromata (MEN 2b) • Bluish sclerae (Marfan) • Lens displacement (up in Marfan, down in homocystinuria) **Visual acuity** • Myopia (Marfan, homocystinuria) **Visual fields** • Bitemporal hemianopia (pituitary tumour) • Homonymous hemianopia (cerebral thrombosis with homocystinuria) **External ocular movements** • Ophthalmoplegia (hyperthyroidism) • Sixth cranial nerve palsy (intracranial tumour) **Cataracts** (homocystinuria)
Nose	Anosmia (Kallmann)
Mouth	**Lips**: prominent (neuromata in MEN 2b)

iv. Arm span

Arm span - Total height: normal values

Birth-7yr = −3 cm	8-12 yr = 0cm	14yr (girls) = +1cm	14yr (boys) = +4 cm

v. Head circumference

vi. Weight

STEP III: MANOEUVRES

I. Hands and feet together

To detect:
- Hemihypertrophy (Beckwith, McCune–Albright, Proteus)
- Unilateral growth arrest (homocystinuria with cerebrovascular accident)
- Genu valgum (homocystinuria)
- Genu recurvatum (Marfan)
- Pes planus (Marfan)

2. Bend over and touch toes

To detect:
- Scoliosis (Marfan, homocystinuria, Proteus, Sotos, NF-1)
- Kyphosis (with scoliosis, as above, pituitary gigantism)

3. Beighton score for hypermobility

i) Put palms flat on the floor, while reaching down/bending forward, without any bending of the knees
ii) apposing thumb to forearm with wrist flexed
iii) passive hyperextension of the fifth finger over 90° (Gorling's sign)
iv) hyperextension of knees greater than or equal to 10° (genu recurvatum)
v) more than 10° hyperextension of the elbows (Not useful in Marfan)
If these tests show less mobility than normal, then homocystinuria is more likely.

4. Arachnodactyly tests: Steinberg (thumb past ulnar border)/Walker–Murdoch (wrist)

Test for arachnodactyly (Marfan), using two digit-related eponymous signs:
(a) the thumb (Steinberg) sign—this is an extension of the whole distal phalanx of the thumb beyond the ulnar border of the hand when apposed across the palm;
(b) the wrist (Walker–Murdoch) sign—this is overlapping of the distal phalanx of the thumb with the distal phalanx of the little finger when encircling the opposite wrist.
If both these tests are normal, then Marfan syndrome is a less likely diagnosis.

5. Arms out straight

Finally, ask the child to hold the arms out straight in front with the fingers spread apart and check for tremor associated with hyperthyroidism.

*If the patient appears to be quite clearly Marfanoid, the examination outlined can be substantially abbreviated and can concentrate on the **skeleton, eyes** and **heart**.*

STEP IV: SYSTEMATIC RELEVANT EXAMINATION

Gait	• Shuffling gait; like Chaplin's Little Tramp (homocystinuria) • Hemiplegic gait (homocystinuria with CVA)
Upper limbs	**Arachnodactyly** (Marfan, MEN 2b) **Large hands** (Sotos, Proteus, pituitary gigantism) **Nails**: thyroid acropathy (hyperthyroidism)

TALL STATURE

1) General Look
2) Measurements
3) Manoeuvres
4) Systematic relevant examination

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Ask *'Kis class main parhtaay hain?'*
Impression of mental state: intellectual impairment (homocystinuria, Sotos, Beckwith–Wiedemann [B–W], Klinefelter)

Body habitus
- Marfanoid (dolichostenomelia: Marfan, homocystinuria, MEN 2b)
- Eunuchoid (Kallman, Klinefelter)

Tanner staging
- Delayed (Kallmann, Klinefelter)
- Advanced (precocious puberty, virilisation, e.g. adrenal tumour, CAH)

Skeletal anomalies
- Asymmetry (Beckwith, NF-1, McCune–Albright [McC–A], Proteus)
- Pectus excavatum (Marfan, homocystinuria, MEN 2b)
- Scoliosis (Marfan, Sotos, homocystinuria, MEN 2b, NF-1)

Posture
- Hemiplegic (homocystinuria)

Skin
- Café-au-lait spots (NF-1, McC–A, Proteus)
- Subcutaneous tumours, e.g. lipomata, haemangiomata (Proteus)
- Hyperpigmented areas (McC–A, Proteus)
- Acne (virilisation syndromes)

STEP II: MEASUREMENTS

i. Standing height
ii. Upper segment (US) (Sitting height)
iii. Lower segment (LS) is calculated by subtracting US from total height

U/L ratio normal value

Birth = 1.7	3 yr = 1.3	8yr = 1	18yr = 0.9

INVESTIGATIONS

Aj's Super-table for Short stature workup

Step1

Height >2 to <5th percentile Following own growth curve Has family history of short stature/pubertal delay		Bone age CBC Chemistry profile ESR UA	± Celiac panel ± Free T4/TSH ± IGF-1 and IGFBP3(GH) ± Karyotype (Turner) ± CF, HIV	Step2
Bone age				
Family history of short stature	Family history of pubertal delay	Weight < height for age	Weight ≥ height for age	
Normal puberty	Pubertal delay			
Bone age = chronological age	Bone age < chronological age Bone age = height age	**Chronic illnesses, Malnutrition, Malabsorption**	U/L DisPro-portionate	U/L Pro-portionate
Familial short stature	**Constitutional growth delay**		**Skeletal Dysplasia**	**Endocrine**

** Insulin-like growth factor (IGF)-1 and IGF binding protein 3 (IGFBP3)*

Bone age is most useful

In girls, a chromosomal analysis is always warranted to exclude Turner syndrome.

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELLING

III. SPECIFIC TREATMENT

Treat the cause

NOTES

TURNER SYNDROME DESCRIPTION

Manoeuvres for short stature revealed

1) Wide spaced nipples
2) Shield like chest
3) Short fourth metacarpal
4) Increased carrying angle
5) Neck webbing
6) Low hairline

GH DEFICIENCY DESCRIPTION

Examination revealed

1) Infantile/ Cherubic facies
2) Infantile voice
3) Midline defects (cleft palate, single central maxillary incisor)
4) Central obesity
5) Micro penis
6) Cryptorchidism/undescended testis

ACHONDROPLASIA DESCRIPTION

1) Disproportionate short stature
2) Short arm span
3) Rhizomelic/mesomelic shortening
4) Spinal deformities (lordosis/kyphosis)

DIFFERENTIALS

PROPORTIONATE SHORT STATURE	DISPROPORTIONATE SHORT STATURE
Constitutional **Familial** **Malnutrition** **Chronic illnesses** **Syndromes** **Psycosocial deprivation** **Endocrinopathies** **Osteogenesis imperfecta**	**Skeletal dysplasia except OI** **Congenital Hypothyroidism** **Rickets**
Labs: Bone age CBC, ESR,RFTS, anti-TTG Hormonal study	Labs: Skeletal survey TFTs Calcium profile

BONE AGE X RAYS

Birth : Knee	<7 yr : Wrist	>8 yr : Elbow

STEP IV: SYSTEMATIC RELEVANT EXAMINATION

Sitting on couch

General	Nutritional status, Hyperpigmentation (cushing) Diagnostic facies *(Turner syndrome, Down syndrome, Klinefelter)* Obesity (*Prader-Willi / Laurence-Moon-Biedl syndromes, Cushing*)
CNS	Focal neurological signs
Hands	Short stubby hands, poly or syndactyly, clubbing, pallor, wrist widening
Arm	Radial pulse, epitrochlear & axillary lymph nodes, BCG scar
Neck	JVP, Thyroid exam, Cervical lymph nodes
Hair & Scalp	Ant. Fontanelle (wide open), Scalp hair
Eyes	Jaundice, pallor, bitot spots
Mouth	Repaired cleft lip or palate, Dental assessment

Lying on couch

Chest	Auscultate heart timing with carotids, Auscultate chest(CF, asthma), deformity
Abdomen	Liver, spleen, kidneys, Protuberant abdomen
Genitalia	Seek permission! … for SMR
Legs	Pedal edema
Vitals & GPE	Apply cuff for Blood pressure (Renal disease, Cushing)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Child's progressive percentile charts (for growth velocity)
3) Parents' heights and onsets of puberty (for MPH & constitutional delay)
4) Visual field examination (for craniopharyngioma)
5) Fundoscopy (for papilledema/ craniopharyngioma)
6) Child's X ray wrist (for Bone age)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
Her/his **total height** is _____cm (which is below 3rd centile for his/her age and sex) with **proportionate/disproportionate Upper & lower segments.**
His **weight** is _____kg & **Fronto-occipital circumference** is _____cm (but I will like to plot it on centile charts)
His/her **Vitals** are _____
Manoeuvres for short stature revealed no abnormality/abnormal _____
There are No signs of **hypothyroidism, micronutrient deficiency, edema or lymphadenopathy.**
There is no clinical **evidence of chronic heart, lung, liver or renal disease.**
(Describe skeletal deformity in case of disproportionate short stature)
BCG scar is seen.**SMR** is prepubertal.

5. ARMS OUT STRAIGHT

Cubitus valgus (Turner, Noonan)
Over 15° in girls;
Over 10° in boys
Restriction of elbow extension (e.g. hypochondroplasia)

II. INSPECT FROM THE SIDE (standing with the arms by side)

Head & Neck

1) Prominent forehead (e.g. achondroplasia)
2) Flat occiput (Down)
3) Proptosis (e.g. syndromes with craniosynostosis)
4) Micrognathia (e.g. Pierre Robin sequence)
5) Prognathism (e.g. achondroplasia)

Upper limbs

1) Short upper limbs : fingers may only reach the proximal thigh
2) Short trunk : fingers may reach the knees

Shape of back

1) Lordosis (e.g. achondroplasia),
2) Thoracolumbar kyphosis (e.g. achondroplasia)
3) Crouched posture (e.g. diastrophic dysplasia)

III. INSPECT FROM THE BACK

1. BACK

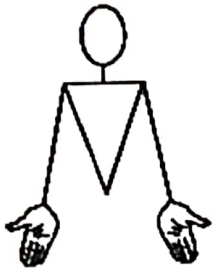
Short neck (Klippel–Feil, Noonan)
Neck webbing (Turner, Noonan)
Low hairline (Turner, Noonan, Klippel–Feil)

2. BEND OVER AND TOUCH TOES (to determine structural scoliosis)

Scoliosis (e.g. Noonan, Klippel–Feil, Prader–Willi, trisomies)
Wasted buttocks

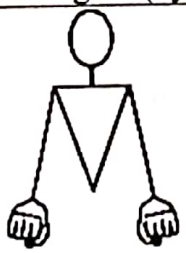
1. HANDS AND FEET TOGETHER

Asymmetry (Russell–Silver)
Approximation of shoulders (absent clavicles in cleidocranial dysostosis) **feel & see clavicles**



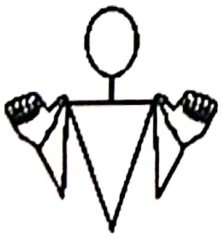
2. PALMS UP

Simian crease (Down, Seckel)
Clinodactyly (Russell–Silver, Down, Seckel) (Clino=curved)
Short fingers (hypochondroplasia), Syndactyly (e.g.Apert syndrome)



3. MAKE A FIST

Short fourth metacarpal
(pseudohypoparathyroidism, Turner, fetal alcohol)
Trident deformity in achondroplasia
Check the nails (e.g. hyperconvex in Turner)



4. THUMBS ON SHOULDERS

Proximal segment shortening (e.g. achondroplasia, hypochondroplasia)
Middle segment shortening (e.g. Len–Weill dyschondrostenosis, Langer mesomelic dysplasia)
Distal segment shortening (e.g. acromesomelic\dysplasia)
Thumbs overshoots : Proximal segment (rhizomelic) limb shortening
Thumbs do not reach the shoulders: middle segment (mesomelic) or distal segment (acromelic) limb shortening, or alternatively the limbs may be bent (camptomelic).

SHORT STATURE

1) General Look
2) Measurements
3) Manoeuvres
4) Systematic relevant examination

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Listen carefully to the patient's age
Stand back & look for any evidence of an obvious diagnosis (e.g. Turner or Noonan syndromes)
Dysmorphism and any disproportion (skeletal dysplasias, rickets)
Nutritional status
Pubertal status (visually assess)
Delayed puberty = constitutional delay, pituitary disorders and chronic diseases
Normal puberty = familial short stature

STEP II: MEASUREMENTS

i. Standing height
ii. Upper segment (US) (Sitting height)
iii. Lower segment (LS) is calculated by subtracting US from total height

U/L ratio normal value

Birth = 1.7	3 yr = 1.3	8yr = 1	18yr = 0.9

iv. Arm span

Arm span - Total height: normal values

Birth-7yr = −3 cm	8-12 yr = 0cm	14yr (girls) = +1cm	14yr (boys) = +4 cm

v. Head circumference
vi. Weight

STEP III: MANOEUVRES

I. INSPECT FROM FRONT (stand opposite the child and demonstrate)

INVESTIGATIONS

1. Establish precocious puberty (Increased sex steroids)

Boys	Girls
Mid night testosterone level of pubertal value	Post breakfast estrogen level of pubertal value
Bone age advanced (X-ray wrist)	Bone age advanced (X-ray wrist)

2. Is it Central or Peripheral?

LH, FSH Levels (increased in central) Basal & post-GnRH

3. Identify underlying pathology?

Boys	Girls
Central 1. CT Scan/MRI Brain for tumor/mass	**Central** 1. CT Scan/MRI Brain for tumor/mass (obligatory if PP<3years of age)
Peripheral 1. **Other serum androgen levels**: e.g. 17-OH progesterone, DHEAS, androstendione 2. **Urine: steroid profile** (sex/adrenal steroids) 3. **Pelvic & Abdominal USG** for testicular tumor, hepatoblastoma, adrenal masses 4. **DNA study for G protein pathway** (McCune–Albright)	**Peripheral** 1. **Other serum androgen levels**: e.g. 17-OH progesterone, DHEAS, androstendione 2. **Urine: steroid profile** (sex/adrenal steroids) 3. **Pelvic & Abdominal USG** for ovarian tumor, hepatoblastoma, adrenal masses 4. **DNA study for G protein pathway** (McCune–Albright)

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELLING

Explanation of the physiology and physical consequences of precocious puberty
The decision on therapy should be made jointly with the parents

III. SPECIFIC THERAPY

CENTRAL PP

Long acting GnRH analogue SC or IM injection, monthly (or 3-monthly in depot preparations).
Surgery/chemotherapy for tumors
Reduce sex steroid synthesis

CAH

Hormone replacement

IV. MONITORING

Monitoring for Treatment efficacy
1. Growth rate
2. Pubertal stage
3. LH and FSH levels (basal and stimulated)

TIPS & TRICKS

1) Usually a male child is brought as exam case; Command is GPE or Abdomen.
2) Catch is a child taller for his age or having Axillary hair.
3) Increased breast in female OR >4ml testicular volume in male = Central Precocious Puberty

Testicular Volume (*Request orchidometer*), Length of Penis
Abdomen: Skin pinch for hydration status, Mass (Hepatoblastoma, ovarian/adrenal mass)

STEP V: LOWER LIMBS

Reflexes (For UMN signs)
Edema

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Fundoscopy (raised ICP)
3) Serial growth charts (for growth Velocity)
4) Complete Neurological exam

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
There is no **respiratory distress** or **dysmorphism**.
He is **afebrile** with **Respiratory rate** ___/min, **pulse** _____/min, regular in rhythms, normal in volume and character. **BP** is ____mmHg (but I would like to plot it on centile charts)
His **Height** is ____cm, **weight** is ____Kg, **FOC** ____cm (I would like to plot it on centile charts)
There are areas of **skin hyperpigmentation** at _____
He has **axillary & pubic hair, penile length** is____cm with **tanner stage** _____. I would like to check **testicular volume** by orchidometer.
There is no evidence of **pallor, dehydration, lymphadenopathy, CN palsies, abdominal mass, scoliosis** or **edema**.

DIFFERENTIALS

Central (true) PP (gonadotrophin-dependent)	Peripheral PP (gonadotrophin-independent)
1) **Idiopathic** (familial/non-familial) 2) **Intracranial tumours**: e.g. Hypothalamic hamartoma, craniopharyngioma, astrocytoma, optic glioma 3) **Other CNS lesions**: Hydrocephalus, arachnoid cysts, traumatic brain injury, cranial irradiation, hamartomas, post-infective 4) **Secondary central PP**: Early maturation of the hypothalamic–pituitary–gonadal axis due to long-term sex steroid exposure, e.g. CAH, McCune–Albright syndrome.	1) **Gonadal**: McCune–Albright syndrome; ovarian tumours (e.g. benign cyst; granulosa cell tumour); testicular tumour; familial testitoxicosis 2) **Adrenal**: CAH; Adrenal tumour (carcinoma; adenoma) 3) **Human chorionic gonadotrophin (HCG)-secreting tumours**: e.g. CNS (chorioepithelioma; dysgerminoma) Iatrogenic (exogenous sex-steroid administration)

PRECOCIOUS PUBERTY

1) General Look
2) Gait, Anthropometry & Back
3) Relevant GPE
4) Genitalia & Abdomen
5) Lower limbs

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building ***(Talk to child to assess voice)***
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

Listen to Age clearly: Watch for inappropriate physique for age

STEP I: GENERAL LOOK

Dysmorphism (McCune Albright, Russel silver), Moon facies (adrenal pathology), Hirsutism, Muscular growth, Acne, Pigmentation
Respiratory rate

STEP II: GAIT , ANTHROPOMETRY & BACK

Ask: *'Bachaa Chal saktaa hai?'*

Back	Scoliosis, cafe-au-lait spots
Check gait	
Height	Short = Cushing, Hypothyroid, NF-1, Russel silver , Long standing precocious
	Tall = McCune Albright , Recent CAH
	Arm span > Height = Growth spurt
Weight	↑ in Cushing, hypothyroid
FOC	Hydrocephalus, Intra-Cranial tumors
VP shunt	Hydrocephalus, Intra-Cranial tumors

STEP III: RELEVANT GPE (Make child sit on couch)

Hands	Nails, Pulse
BP	Raised in 11 Hydroxylase deficiency, IC tumors
Arm & Axilla	BCG scar, Axillary hair
Eyes	Cranial nerve palsies secondary to IC tumors, Visual acuity/field, Make H
Thyroid	For Goiter

STEP IV: GENITALIA & ABDOMEN

Seek permission to expose breast & genitals for SMR / Tanner Staging

effects of Thyroxine deficiency on bones	**X-ray skull**: Wide pituitary fossa, Lack of outer & inner table differentiation, Silver beaten appearance due to myxedematous tissue **X-ray Chest**: Cardiomegaly **X-ray spine**: Beaking
VII.To find out associations	**CBC** (Pernicious anemia) **ANA** (SLE) **RA factor** (RA) **Serum Cortisol levels** (Addison's disease) **HbA1c** (DM) **Karyotyping** (Down, Turner)

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELLING

Depending upon cause

III. PHARMACOLOGICAL TREATMENT

Hypothyroidism: Tab Levothyroxine

Hyperthyroidism: Propranolol, Antithyroid drugs (Propylthiouracil, Carbimazole)

IV.SURGICAL TREATMENT

For very large goitre

V.RADIOACTIVE TREATMENT

For hyperthyroidism

TIPS & TRICKS

1) Thin & lean child is usually of hyperthyroidism while short & fat child is usually of hypothyroidism.

VIVA

GRADES OF THYROMEGALLY

Grade 0	No goiter
Grade 1a	Goiter palpable but not seen with normal neck position
Grade 1b	Goiter seen after maximum dorsi-flexion of neck
Grade 2	Goiter visible with normal head position
Grade 3	Goitre visible at a distance

ASSOCIATIONS OF GOITER

Turner, Noonan, Down syndrome.

Addison disease, SLE, Pernicious anemia, Rheumatoid arthritis, DM

NOTES

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
Her **voice** is normal / hoarse & there is no **distress** or **dysmorphism**.
She looks **agitated / anxious / depressed**.
She has a **bilobular anterior neck swelling** with no visible **pulsations, scars, prominent veins** or **redness**.
It is **moving with swallowing** but not with tongue protrusion. It is **non-tender, non-pulsatile, diffuse, firm** in consistency with normal **temperature**. It **measures** __ × __cm with palpable **lower margins** & there is no **bruit**.
There is no **cervical lymphadenopathy**.
There was no evidence of **sweating on forehead, miosis, ptosis** and **Pemberton's sign** was absent.
She is **afebrile**; **Respiratory rate** is ___/min; **pulse** is ___/min regular in rhythm & normal in volume & character. Her **B.P.** is ____mmHg. Her **height** is ____ cm; **weight** is ___kg; **FOC** is ___cm but I would like to plot them on centile charts.
Reflexes are normal /delayed and there is no evidence of **tremors, dry / sweaty skin, wide anterior fontanelle, lid lag, retraction, exophthalmos, , pallor (hypothyroidism, pernicious anemia), jaundice (AIH), Murmur, Umbilical hernia, Hepatomegaly** or **edema.**

DIFFERENTIALS

EUTHYROID	THYROTOXICOSIS	HYPOTHYROIDISM (ACQUIRED)
Endemic goiter Simple colloid goiter Euthyroid Hashimoto Subacute thyroiditis Graves' disease on R_x	Graves' disease Hashitoxicosis Subacute thyroiditis	Hypothyroid Hashimoto Iodine deficiency
Normal Vitals & Anthropometry; No Bruit; No Eye signs; No tremors & sweaty hands	Thin, sweaty hands, fine tremors, wide pulse pressure, elevated systolic BP, bruit, ophthalmic signs	Myxedematous facies, Rough, dry skin, hoarse voice, bradycardia, slow relaxation, hypotonia

INVESTIGATIONS

I. To check thyroid status	T_4, TSH (If normal: check thyroid antibodies) TRH estimation (↑=Pituitary disorders; ↓=Hypothalamic disorders)
II. To identify presence of thyroid	USG of thyroid Technetium scan ^{99}Tc (Presence/Absence/Normal/Ectopic)
III.To identify dyshormonogenesis	Radiolabelled Iodine ^{131}I uptake
IV.To identify autoimmune cause	Anti-thyroid antibodies (Anti-microsomial Ab; Antithyroid peroxidase Ab; Anti-thyroglobulin Ab)
V.To Rule out Iodine deficiency	Urinary Iodine excretion
VI.To identify	**X-ray wrist**: Delayed bone age & stippled epiphysis

1) Check Gland (**size, shape, symmetry, surface, consistency, mobility, tenderness**)
2) **Swallow** / sip of water (check for asymmetry in lobes = unilateral mass)
3) Ask to **protrude tongue**
4) Palpate **Cervical lymph nodes**
5) Assess for **tracheal deviation** from back (large goitre)

STEP III: PERCUSSION

Percuss for retro-sternal dullness (large goitre)

STEP IV: AUSCULTATION

Take deep breath & hold (Hold your own breath too)
With bell → auscultate each lobe for bruit (↑ vascularity 2° Graves' disease)
(To differentiate → bruit of artery is louder along line of carotid artery)

STEP V: TO CHECK DIRECT EFFECTS OF GLAND

Horner syndrome (check sweating on forehead, miosis, ptosis)
Pemberton's sign: *Ask to raise arms above head!* (stridor, facial congestion, neck vein distension on raising arms = Retrosternal thyroid)

STEP VI: EYE EXAMINATION

From front	*Look straight on wall behind me* → **lid retraction** (sclera visible above iris)
	Make H-sign (to look for **restriction in eye movement**)
	Bring finger from above to down (**Lid lag**)
	Look for **pallor** of conjunctiva, **jaundice**
From both sides and back	Look for anterior Displacement of eye out of orbit (**exophthalmos**)

STEP VII: RELEVANT GPE

Hands	Bring hand in front, place paper on it (tremor)
Pulse	↑=Hyper; ↓=Hypo; Irregular (AF) = Hyper
Vitals	B.P., Temperature. (if time persists)
Scalp	Anterior Fontanelle (wide open in younger child with Hypothyroidism)
Ask to lie down on couch	
CVS	Murmur
Abdomen	Umbilical hernia, Hepatomegaly (Autoimmune hepatitis)
Reflexes	Knee = ↓Reflexes = Hypothyroidism
Edema	Pre-tibial myxedema (Graves' disease)
Ask to stand	
Gait	Sit up (Gower's sign) (Proximal myopathy = Hyperthyroidism)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) Formal IQ assessment (Congenital Hypothyroidism)

GOITRE

1) Inspection
2) Palpation
3) Percussion
4) Auscultation
5) To check direct effects of gland
6) Eye examination
7) Relevant GPE

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
   1) **Talk to child to check voice** (*recurrent laryngeal nerve paralysis*) + **assess hearing** (*Pendred syndrome*)
   2) **Shake hand to assess for sweating** (both hands)
c. **Position patient [Sitting** on couch/parent's lap]
d. **Exposure:** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: INSPECTION

Check Respiratory Rate (6sec) × 10
Stand back → *ask Patient to extend neck* (give demo) **LOOK, LOOK, LOOK**
1) **What is it? Eu / Hyper / Hypo-thyroid?**

Hyperthyroidism	Hypothyroidism
Agitated / Anxious	Dull look, Hyperpigmentation / dysmorphism (turner, down syndromes)
Thin / lean	Short Statue
Tremors	Facial myxedema
Exophthalmos	Thyroidectomy scar

2) **Look at the neck for goiter & Grade it** (*see in Viva 'Grades of Goitre'*)
Skin changes, scar, any obvious swelling on either side of trachea & on front.
Ask patient to swallow / drink water
Ask patient to protrude tongue. (See tremors)
- No movement (thyroid gland mass / lymph node)
- Upward movement (Thyroglossal cyst)

STEP II: PALPATION (FROM BACK)

Ask about pain in swelling before palpating!
Warm your hands & **Check temperature** of swelling with dorsum of your hand.
Ask Patient to flex his neck & relax → begin with Adam's apple (thyroid cartilage) → Move down to superior edge of cricoid cartilage; (below it is isthmus) palpate isthmus & both lobes.

After age 2 yr	Macrovascular disease	**Lipids**, BP	Every 5 yr	Statins for hyperlipidemia BP control
After 5 yr duration (Prepubertal) After 2 yr duration (Pubertal)	Retinopathy	**Fundal photography**, Fluorescein angiography, mydriatic ophthalmoscopy	Annually	Improved glycemic control, Laser therapy
	Nephropathy	**Spot urine sample for albumin : creatinine ratio** 24 hr excretion of albumin, urinary albumin : creatinine ratio	Annually	Improved glycemic control, BP control, ACE inhibitors
5 yr after diagnosis	Neuropathy	**GPE** Nerve conduction, thermal & vibration threshold,pupillometry, cardiovascular reflexes	Unclear?	Improved glycemic control

PROGNOSIS & COUNSELLING

IDDM affects 1 in 500 children under 18 yrs of age
Siblings/offsprings of diabetic have 5% risk (identical twins 50% risk)

NOTES

	the oral intake is limited	blood glucose <250 mg/dL	from body.

Call physician or nurse:
1) If blood glucose remains elevated after 3 extra doses
2) If blood glucose remains less than 70 mg/dL and child cannot take oral supplement
3) If dehydration occurs

Emergency department visit:
1) Large ketonuria and emesis
2) Blood glucose < 50–60 mg/dL (2.8–3.3 mmol/L) & poor oral intake

MANAGEMENT DURING SURGERY

Intravenous Insulin Coverage During Surgery

BLOOD GLUCOSE LEVEL (mg/dL)	INSULIN INFUSION (units/kg/hr)	BLOOD GLUCOSE MONITORING
<120	0.00	1 hr
121–250	0.03	2 hr
250–300	0.06	2 hr
300–400	0.08	1 hr* (Check urine ketones)
400	0.10	1 hr* (Check urine ketones)

Timing of surgery	When surgery is elective, it is best performed early in the day, allowing the patient maximal recovery time to restart oral intake & subcutaneous insulin therapy.
Fluids during surgery	Insulin infusion + 5% glucose and 0.45% saline solution with 20 mEq/L of potassium acetate is given at 1.5 times maintenance rate.
Brief elective surgery	When elective surgery is brief (less than 1 hr) and full oral intake is expected shortly afterward → Simply monitor the blood glucose hourly + give a dose of insulin analog according to the child's home glucose correction scale.
Basal insulin	If glargine or detemir is used as the basal insulin, a full dose is given the evening before planned surgery.
NPH/Lente	If NPH or Lente is used, one half of the morning dose is given before surgery. The child should not be discharged until blood glucose levels are stable and oral intake is tolerated.
Post OP care	The IV insulin is continued after surgery as the child begins to take oral fluids; the IV fluids can be steadily decreased as oral intake increases. When full oral intake is achieved, the IV may be capped and subcutaneous insulin begun.

VIII. SCREENING AND MANAGING COMPLICATIONS

When to start screening	Disease	Tests	Frequency	Intervention
At diagnosis	Thyroid disease	**TSH** Thyroid peroxidase, thyroglobulin antibodies	Every 2-3 yr	Thyroxine
	Celiac disease	Tissue transglutaminase, Endomysial Ab	Every 2-3 yr	Gluten-free diet

exposure to it, the higher is fraction of HbA1c, expressed as a percentage of total hemoglobin.

Significance of HbA1c

1) Because a blood sample at any given time contains a mixture of red blood cells of varying ages, exposed for varying times to varying blood glucose concentrations, an HbA1c measurement reflects the average blood glucose concentration from the preceding 2-3 mo.
2) When measured by standardized methods to remove labile forms, the fraction of HbA1c is not influenced by an isolated episode of hyperglycemia.
3) The lower the HbA1c level, the more likely it is that microvascular complications such as retinopathy and nephropathy will be less severe, delayed in appearance, or even avoided altogether.

Frequency of measuring HbA1c

3-4 times/yr to obtain a profile of long-term glycemic control

Values of HbA1c

Non-diabetic <6%	Diabetic ≥6.5%	Target HbA1c <7.5%

Good control	Fair control	Poor control
6-7.5%	7.6-9.9%	≥10%

Target Pre-meal BSR & HbA_{1c}

Age group (yr)	**Pre-meal BSR** (mg/dL)	**HbA_{1c}** (%)
<5	100-**200**	7.5-**9.0**
5-11	80-**150**	6.5-**8.0**
12-15	80-**130**	6.0-**7.5**
16-18	70-**120**	5.5-**7.0**

Non-diabetic reference range for HbA1c is 4.5-5.7%

VII) SPECIAL SITUATIONS

MANAGEMENT DURING INFECTIONS/ SICK DAY GUIDELINES

URINE KETONE STATUS	GLUCOSE TESTING AND EXTRA RAPID-ACTING INSULIN		COMMENT
	Insulin	Correction Doses	
Negative or small	q2hr	q2hr for glucose >250 mg/dL	Check ketones every other void
Moderate to large	q1hr	q1hr for glucose >250 mg/dL	Check ketones each void, Go to hospital if emesis occurs

Mnemonic: **SICK**

Sugar	**I**nsulin to prevent DKA	**C**arbs	**K**etone
Check blood glucose more frequently (2-3 hrly)	**Never Stop Insulin** Basal insulin: (glargine/detemir) should be given at the usual dose and time. NPH / Lente : Reduced by half if blood glucose <150 mg/dL and	Make sure to take enough carbohydrates & drink enough fluid **Oral fluids** Sugar-free if blood glucose >250 mg/dL (14 mmol/L); Sugar-containing if	Frequently check urine/blood ketones (4hrly) Use extra rapid acting insulin if ketones are present Drink plenty of fluid to flush out ketones

IV) DIET

Nutrient	Calories	Recommended daily intake
Carbohydrate	55%	70% of the carbohydrate should be derived from complex carbohydrates such as starch. (require prolonged digestion & absorption so that plasma glucose levels increase slowly) Limit intake of sucrose and highly refined sugars (rapidly absorbed and may cause wide swings in metabolic pattern)
Protein	15%	
Fat	<30%	
	Saturated: <10%	Polyunsaturated: 10% — Monounsaturated: Remainder of fat allowance
	Polyunsaturated:saturated ratio is increased to approximately 1.2 :1.0	
Fiber	>20 g/day	High fiber, especially soluble fiber: optimal amount?
Cholesterol	300 mg	Limit number of egg yolks consumed. Simple measures reduce serum low-density lipoprotein cholesterol, a predisposing factor to atherosclerotic disease
Sodium		Avoid excessive: 3,000–4,000 mg if hypertensive

V) BASIC & ADVANCED DIABETES EDUCATION

1) In the acute phase, the family must learn the "basics," which includes monitoring the child's blood glucose and urine and/or blood ketones, preparing and injecting the correct insulin dose subcutaneously at the proper time, recognizing and treating low blood glucose reactions, and having a basic meal plan.
2) Written materials covering these basic topics help the family during the 1st few days.
3) Children & their families are required to complete advanced self-management classes in order to facilitate implementation of flexible insulin management (during athletic activities/sick days)

VI) MONITORING

Glucose oxidase strips	Daily blood glucose monitoring has been markedly enhanced by availability of strips impregnated with glucose oxidase that permit blood glucose measurement from a drop of blood
Frequency of monitoring	4 times daily—**before breakfast, lunch, and supper, and at bedtime** When insulin therapy is initiated/adjustments are made that may affect the overnight glucose levels, self-monitoring of blood glucose should also be performed at **12 midnight and 3 am** to detect nocturnal hypoglycemia. **More frequent** blood glucose monitoring : based on level of physical activity and history of frequent hypoglycemic reactions
Range for BSR	**80 mg/dL** in the fasting state to **140 mg/dL** after meals
Glycosylated Hemoglobin	A reliable index of long-term glycemic control. **Introduction** 1) HbA1c represents the fraction of hemoglobin to which glucose has been non-enzymatically attached in the bloodstream. 2) The formation of HbA1c is a slow reaction that is dependent on the prevailing concentration of blood glucose; it continues irreversibly throughout the RBC's life span of 120 days. 3) The higher the blood glucose concentration and the longer the RBC's

3) ABGs (to exclude DKA, low pH, Bicarbonate base deficit↓, total CO_2)
4) Anion gap increased
5) CBC ↑ Hb & Hct due to dehydration, ↑ WBCs)
6) Serum osmolality elevated
7) Urea, creatinine elevated
8) Neuroimaging (cerebral edema)

III. TO RULE OUT COMPLICATIONS

1) Testing for autoimmunity

Testing for autoimmunity to β cells	Not commonly necessary in the non obese child Islet cell antibody (ICA) Anti-insulin antibody (IAA)/ Anti-IA-2 Anti-GAD antibody
Testing for other autoimmune diseases	Antithyroid peroxidase and antithyroglobulin antibodies (hypothyroidism) (↑ in 15-30%) T4, TSH should be checked after the child is stable for a few weeks as significant physiologic distress can disrupt the pituitary–thyroid axis Tissue transglutaminase IgA and total IgA (celiac disease) (in 5-10%)

2) Testing for long term complications

Fundal photography/ ophthalmoscopy	(for Retinopathy) 1-2 yearly
Spot urine for albumin : creatinine ratio	(for Nephropathy) Annually
Lipid profile	(for Macrovascular disease) 5 yearly

TREATMENT

a) Tabulated Overview

Mnemonic: Exercise DIDM–Exercise, Insulin, Diet, eDucation, Monitoring & Follow up

I)MDT	II)Exercise	III)Insulin	IV)Diet	V)Education	VI)Monitoring & Follow up
VII) Special situations		VIII) Screening and Managing complications		

b) Details

I) MDT (Multi-disciplinary team approach)

Pediatrician	Endocrinologist	Ophthalmologist	Nephrologist
Neurophysician	Dietician	Psychologist	Psychiatrist

II) EXERCISE

1) At least 25 min regular aerobic exercise (↓insulin requirement)
2) No form of exercise, including competitive sports, should be forbidden to the child with diabetes.

III) INSULIN

Starting Doses of Insulin (units/kg/day)

	NO DKA	DKA
Prepubertal	0.25-0.50	0.75-1.0
Pubertal	0.50-0.75	1.0-1.2
Postpubertal	0.25-0.50	0.8-1.0

Thyroid was not enlarged
Abdomen had multiple injection sites without atrophy But there was no distention or visceromegaly.
Tanner staging was pre-pubertal & there was no evidence of **atrophy, infection** or **peripheral neuropathy** on examining lower limbs.
Skin was Not hyperpigmented & breath sounds were vesicular.

INVESTIGATIONS

a) Tabulated Overview

I. FOR DIAGNOSIS	II. TO R/O DKA	III. TO RULE OUT COMPLICATIONS
1) BSR 2) BSF 3) Post prandial BSR 4) Hb A_{1C} 5) Urine for glucose	1) Urine for ketones 2) Serum electrolytes 3) ABGs 4) Anion gap 5) CBC 6) Serum osmolality 7) Urea, creatinine 8) Neuroimaging	1) Testing for autoimmunity Autoantibodies Celiac screen Thyroid screen 2) Testing for long term complications Fundoscopy Spot urine for albumin : creatinine ratio Lipid profile

b) Details

I. DIAGNOSTIC CRITERIA (Confirming hyperglycemia)

	IMPAIRED GLUCOSE TOLERANCE	DIABETES MELLITUS
Random plasma glucose		Polyuria, polydipsia, & unexplained weight loss with glucosuria ± ketonuria + **BSR ≥200 mg/dL (11.1 mmol/L)**
Fasting plasma glucose (≥8hr)	**100-125 mg/ dL** (5.6-7.0 mmol/L)	**≥126 mg/dL** (7.0 mmol/L)
2-hr plasma glucose (OGTT)	**≥140 mg/dL, but <200 mg/dL (11.1 mmol/L)**	**≥200 mg/dL**
Initially post prandial hyperglycemia then fasting hyperglycemia develops		
Hb A_{1C}		**≥6.5%**

A baseline hemoglobin A_{1c} allows estimate of the duration of hyperglycemia & provides an initial value by which to compare the effectiveness of subsequent therapy.

Spuriously elevated HbA1c	Spuriously lower HbA1c
Thalassemia **Other conditions with elevated hemoglobin F** (e.g. hereditary persistence of HbF, hereditary spherocytosis, aplastic anemia, thyrotoxicosis)	**Sickle cell disease** **Other conditions with high red blood cell turnover** (e.g. Hypersplenism, Hemolytic anemia, Hemorrhage, Pregnancy, Pure red cell aplasia, Blood transfusions, Anemias associated with cirrhosis, myelodysplasias, or renal disease treated with EPO)

II. TO DETERMINE DKA (if ketonuria +) **& ITS COMPLICATIONS**

1) **Urine for ketones** (large amount of ketonuria)
2) **Serum electrolytes** (**All Low**: Hyponatremia, Hypokalemia, Low chloride)

	Visual acuity, Eye movements (Neuropathy) Pupillary reactions
Mouth	Hydration, Ketotic breath, Oral candidiasis
Thyroid	Inspect, Swallowing, Palpate, Auscultation (Autoimmune, Thalassaemia)

STEP IV: ABDOMEN

Abdomen	Injection sites, Fat atrophy, hypertrophy Distension (coeliac disease) Hepatomegaly (from glycogen if 'overinsulinised'/fat if 'underinsulinised') Splenomegaly (Thalassaemia)
'Sir, I would like to examine genitalia for tanner staging & perineal candidiasis'	
Genitalia	Tanner staging, Perineal candidiasis (girls)

STEP V: NEUROLOGICAL EXAM

Lower limbs	Injection sites, Fat atrophy, hypertrophy, Trophic changes (thighs) Candidiasis, Necrobiosis lipoidica (lower legs) Reflexes (peripheral neuropathy) Sensation for peripheral neuropathy (Light touch, Vibration, position)

STEP VI: BLOOD PRESSURE

Apply cuff for B.P.	
Blood pressure	Hypertension (nephropathy), Hypotension (Addison) Postural hypotension (autonomic neuropathy, dehydration)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Visual acuity
2) Fundoscopy for Red reflex (cataracts), Retinopathy, Optic atrophy (DIDMOAD)
3) Otoscopy to rule out Ear infection
4) Hearing assessment (for DIDMOAD)
5) Urinalysis (for Glucose, Ketones, Protein & Blood)
6) Request insulin dosages and glucometer readings

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
There is no **distress** or **dysmorphism.**
His/her **HR** is ____/min, **R/R** is ___/min, **B.P.** is _____mmHg and he/she is **afebrile.**
Height is ____cm, **FOC** is ____cm while **weight** is ____kg but I would like to plot on centile charts.
Examination of Back & **Gait** was unremarkable.
Finger prick marks were seen on finger tips of both hands BUT there was no evidence of **Trophic changes , Cutaneous infections** or **palmar pigmentation.**
Cathedral sign was negative.
Eye examination revealed normal Eye movements & Pupillary reactions

DIABETES MELLITUS

1) General Look
2) Anthropometry , Gait & Back
3) Relevant GPE
4) Abdomen
5) Neurological exam
6) Blood pressure

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Face	Scan for thalassaemic facies and hyperpigmentation (Thalassaemia) Clubbing and cough (CF), Friedreich's ataxia, Cushing's syndrome, Wolfram's syndrome/**DIDMOAD**: **D**iabetes Insipidus, **D**iabetes Mellitus, **O**ptic **A**trophy, **D**eafness
Skin	**Hyper-pigmentation** (Thalassaemia), **Hydration**
Respiratory rate	For 10 sec

STEP II: ANTHROPOMETRY, GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

Back	Skin , auscultate chest
Gait	Peripheral neuropathy
Anthropometry	Weight, Height, FOC

STEP III: RELEVANT GPE

Hands	**Fingertip pricks, Trophic changes, Cutaneous infections** **Prayer position** (for Limitation of joint mobility) (looking for lack of apposition of palmar aspects of the fingers (if positive = **cathedral sign**) (If cathedral sign positive : extend DIP,PIP, MCP, Wrist, Elbow)

DIP	extend to 180°	Wrist	extend to 70°
PIP	extend to 180°	Elbow	extend to 180°
MCP	extend to 60°		

Pigmented palmar creases (Addison, thalassaemia with haemosiderosis)

Pulse	Heart rate
Eyes	Squint, Cataract, Contact lenses

VIVA

MEASURES OF TOTAL BODY FATNESS

BMI-for-age percentile charts	Body mass index (BMI) = weight in kilograms/height in metres squared
Waist circumference	Measured at the narrowest [or mid-]point between the lower costal margin and the iliac crest): correlates with abdominal fat and with cardiovascular risk factors (National waist-circumference-for-age percentile charts)
Waist to- height ratio (WHtR)	Aim : Keep the waist to less than half the height

NOTES

He has **truncal obesity** with thin lean extremities. There is **facial plethora, fullness of supra-clavicular fossa** and **pink striae** over abdomen.
There is no evidence of **proximal myopathy, gait abnormality, cataract, nystgmus, bruise, palmar erythema, coarse dry skin, hyperpigmentation, goiter, gynaecomastia** or any obvious **skeletal deformity.**
Abdominal, Cardiovascular and **lower limb** examinations are unremarkable.

DIFFERENTIALS

OBESITY WITH SHORT STATURE	OBESITY WITH N/TALL STATURE
1) Cushing's syndrome 2) Hypothyroidism 3) GH deficiency 4) Prader-Willi syndrome	1) Simple obesity 2) Hypothyroidism (on treatment) 3) GH deficiency (on treatment) 4) Klienfelter syndrome

INVESTIGATIONS

I. SUPPORTIVE
1) **Lipid profile**
2) **LFTs, RFTs, Electrolytes**
3) **BSR, OGTT**
4) **USG Abdomen** (fatty liver, gall stones)
5) **X-ray** for osteoporosis and bone age

II. TO ASCERTAIN CAUSE
1) **Cushing's syndrome:** Cortisol levels, Dexamethasone suppression, ACTH stimulation tests
2) **Hypothyroidism**: T_4, TSH
3) **GH deficiency**: IGF-1, Basal & post stimulation GH levels

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELING
Depending upon cause

III. TREATMENT OF ACUTE PROBLEMS
Anti-hypertensives

IV. SPECIFIC TREATMENT
Simple Obesity: Life style modifications, Exercise, Bariatric surgery
Cushing's syndrome: Transsphenoidal pituitary microsurgery/ Adrenalectomy/ Remove offending drug
Hypothyroidism: Thyroxine
GH deficiency: Growth hormone replacement

TIPS & TRICKS

1) Obesity + Short stature =Pathological obesity (e.g. Cushing's syndrome)
   Obesity +Tall stature= Simple Obesity/ Klinefelter
2) BMI>30 = Obese , BMI>25 = Overweight
3) Must keep BMI charts with you for this case especially if it is part of long case.

	Hypotonic (PW) **Flap** (CO_2 retention)
Pulse	Slow (hypothyroidism), Bounding (CO_2 retention)
BP***	Hypertension (Cushing's, or complication of obesity) (V.V.Important***
Arm	Hold arms up against resistance (proximal myopathy in Cushing)
Head & Neck	**Face** (Xanthomas) **Eyes** (acuity, fields, lens, retina) **Nose** (Anosmia in Kallmann) **Mouth and chin** [Triangular upper lip (PW), Central cyanosis (Pickwickian); Midline defects associated with hypopituitarism or Kallmann] **Delayed dentition** (hypothyroidism) **Oral candidiasis** (Cushing) **Tonsillar size** (upper airway obstruction associated with hypoxia and hypercapnia in Pickwickian) **Micrognathia** (PW) **Facial hair or acne** (Cushing)
Thyroid	**Goitre** (hypothyroidism)

STEP IV: CVS, ABDOMEN & LOWER LIMBS

CVS	For evidence of cor pulmonale (RVH, Loud P2)
Abdomen	Striae, Hepatomegaly (RVF), Adrenal mass
'Sir, I would like to examine genitalia for tanner staging'	
Genitalia	Tanner staging
Lower limbs	**Inspect** : Small feet (PW); Skin for striae, bruises, poor wound healing (Cushing syndrome); External rotation at hip (SCFE) **Measure**: limb lengths for shortening (with SCFE/AVN femoral head) **Palpate** : Ankle oedema (RVF in Pickwickian) **Hip examination**: limitation of internal rotation or abduction (SCFE) **Ankle jerks**: delayed relaxation (hypothyroidism)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Visual acuity
2) Fundoscopy for Retinopathy
3) Hearing assessment (Impaired in Alstrōm's, Kallmann's)
4) Urinalysis (Glucose (PW, Alstrōm's, Cushing's)
5) Progressive percentile charts of child
6) Parents' percentiles

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place
There is no **distress** or **dysmorphism**.
He is afebrile; **pulse** ___ /min regular in rhythm and normal in volume and character, **B.P** ___ **mm Hg; height** is ____ cm; **weight** is ___kg but I would like to plot them on centile charts

CUSHING'S SYNDROME/OBESITY

1) General Look
2) Anthropometry , Gait & Back
3) GPE & BP
4) CVS, Abdomen & Lower limbs

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building *(Mentally retarded=PW,LMBB, Hypothyroid)*
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Face	Dysmorphic features (PW, B–B), Cushingoid features
Skin	Hirsutism, Acne (PCOS); Bruising, Striae (Cushing); Café-au-lait spots (McCune–Albright syndrome)
Distribution of obesity	Central (Cushing)
Visual impression of Tanner Staging	Pseudoprecocity (Cushing) Hypogonadism (syndromes: PW, B–B, Kallmann, Fröhlich) Gynaecomastia (Klinefelter)
Respiratory rate	Hypoventilation (Pickwickian syndrome)

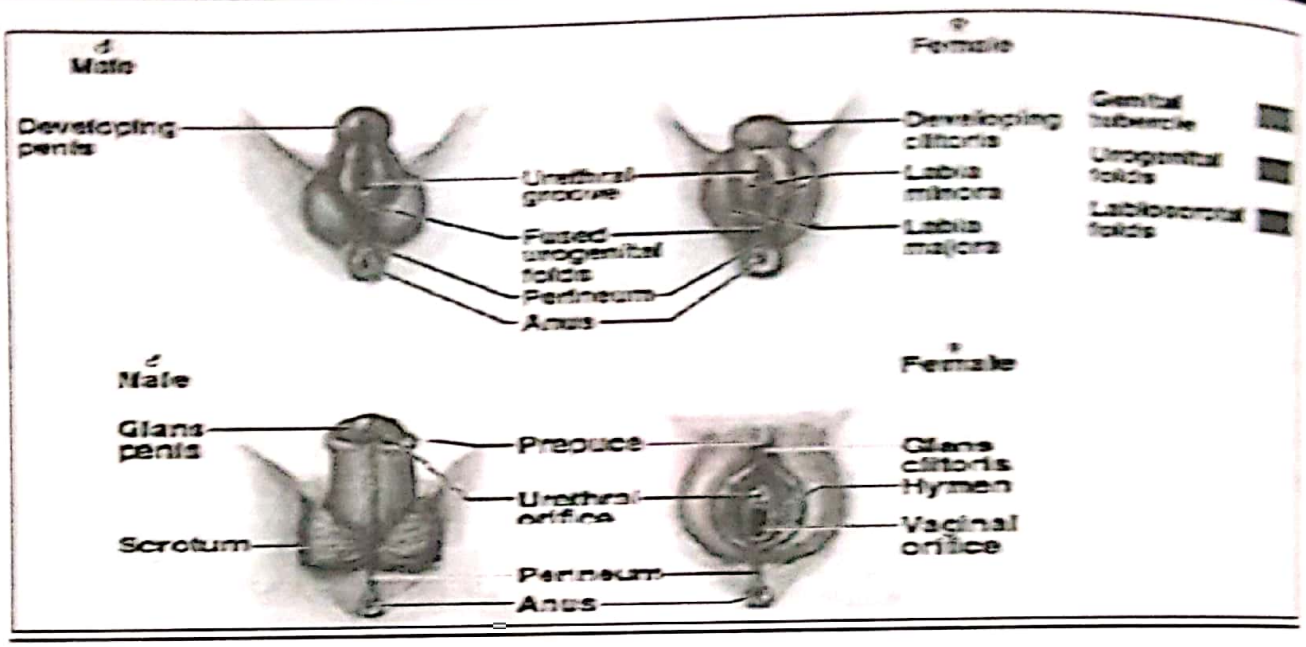
STEP II: ANTHROPOMETRY, GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

Standing with legs together	Look for Leg shortening, External rotation, Genu valgus/varus
Back	**Inspect** 'Buffalo hump', i.e. interscapular fat pad (Cushing's) ; Kyphosis, Scoliosis (Cushing's); Midline scar of MMC repair **Palpate for Spinal tenderness** (Cushing's osteoporosis) **Auscultate for basal crepts** (RVF)
Squat	Proximal myopathy
Gait	Limp in AVN femoral head (Cushing), SCFE (complication of obesity)
Anthropometry	Weight, Height, FOC

STEP III: GPE & BP

Hands	Small (PW)	Short fourth metacarpal (PHPT)
	Polydactyly (B–B)	Scars from removal of additional digit (B–B)
	Temperature	
	Cool (hypothyroidism)	Warm (CO_2 retention in Pickwickian)



NOTES

e.g. significant defects in caudal embryogenesis → complete aplasia of the genital tubercle → absent penis/clitoris
Often, there are ano-genital malformations, not just genital ones.

EMBRYOLOGY

Until 7 weeks' gestation	Bipotential internal genital tracts (in both XX and XY embryos) Genital ducts: Wolffian (mesonephric) & Müllerian (paramesonephric)

SRY (sex-determining region on the Y chromosome) GENE PRESENT (normal male)

Result	Indifferent/ambisexual gonad will develop into a testis *Wolffian duct transforms into epididymis, vas deferens & seminal vesicles.*
8 weeks' gestation	**Sertoli cells** (Testis) : Produce müllerian-inhibiting substance/anti-müllerian hormone (MIS/AMH) **Leydig cells** : Produce testosterone + INSL3(Peptide hormone involved in the regulation of testicular descent)
8 - 12 weeks gestation	1) The müllerian duct regresses due to MIS/AMH. 2) Androgens transform genital primordium into normal male external genitalia. 3) Testosterone is converted by 5-alpha-reductase into DHT
12 weeks until full term	Enlargement of the phallus to normal penile size occurs *All external virilisation is due to androgens*

SRY GENE ABSENT (normal female)

Result	Bipotential gonad will develop into an ovary Müllerian duct → Uterus, fallopian tubes & upper portion of vagina
8 - 12 weeks gestation	Lack of testosterone and MIS/AMH leads to regression of the wolffian duct and preservation of the müllerian ducts.
15 - 25 weeks' gestation	Sexual differentiation of the brain occurs

GENITAL DUCTS - DIFFERENTIATION

	Male (requires SRY)	Female (absence of SRY)
Wolffian duct	Epididymis, vas deferens & seminal vesicles	Regresses
Müllerian duct	Regresses	Uterus, fallopian tubes & upper portion of vagina

Man is a **Wolf**= retains ***Wolffian duct,*** Woman is **Mule** = retains ***Müllerian duct***

EXTERNAL GENITALIA - DIFFERENTIATION

	Male (requires DHT)	Female (absence of DHT)
Genital tubercle	Glans penis	Glans clitoris
Labioscrotal fold	Scrotum (folds fuse)	Labia majora (remain unfused)
Urethral fold	Urethra	Labia minora
Urethral plate		Short female urethra
Urogenital sinus	Bladder and the prostatic urethra	Posterior wall canalizes to form the lower vagina

VIVA

PRADER GRADING OF VIRILISATION OF EXTERNAL GENITALIA

Prader 0: Normal female
Prader 1: Enlargement of phallus (like clitoris; abnormal androgen exposure beyond 8 weeks' gestation)
Prader 2: Enlargement of phallus + Separate vagina and urethra openings
Prader 3: Enlargement of phallus + Single opening (urogenital sinus)
Prader 4: Enlarged phallus + Hypospadias
Prader 5: Normal male

AREA INSPECTED	FINDINGS	POSSIBILITIES
Scrotum	Fused, absent gonads	Must exclude XX with CAH
	Bifid, bilateral gonads	Undervirilised XY True hermaphrodite with bilateral ovotestes (rare)
	Bifid, maldeveloped, bilateral gonads placed high	Undervirilised XY True hermaphrodite with ovotestes (rare) XY gonadal dysgenesis [dysplastic testes]
Midline cleft/urogenital sinus	Gently open cleft/sinus; Locate the urethral meatus Confirm impression of Prader stage (single opening/separate opening) *Skin tags with purplish tinge imply hymen present.*	
Palpation for gonads	Bilaterally palpable, can be brought to base of scrotum	Testes (certainly male) Bilateral ovotestes in true hermaphrodite (rare)
	Bilaterally palpable, but placed high	Undervirilised XY True hermaphrodite with ovotestes (rarely) XY gonadal dysgenesis [dysplastic testes]
	Bilaterally palpable, asymmetrical descent	True hermaphrodite (Testis + Ovotestis) Mixed gonadal dysgenesis (Testis+streak ovary+ hernia)
	Single gonad palpable	True hermaphrodite (Testis + Ovotestis) Mixed gonadal dysgenesis (Testis+streak ovary)
	Impalpable bilaterally	Unpredictable

FIVE GROUPS TO CONSIDER

1. **46,XX DSD**—genetic females with ambiguous or male phenotype.
2. **46,XY DSD**—genetic males with ambiguous or female phenotype.
3. **Ovotesticular DSD**—usually 46,XX (gonads contain both ovarian & testicular components)
4. **Mixed gonadal dysgenesis** (Sex chromosome aneuploidy DSD—for example, 45,X/46,XY mosaicism, with one streak gonad and one dysgenetic testis)
5. **Other**—dysmorphic syndromes, cloacal anomalies, bladder exstrophy etc.

MISTAKENLY AMBIGUOUS (NOT INTERSEX)

1. Premature female infants with an unusual genital appearance (Transient abnormality)
2. Abnormal perineal anatomy (Not due to any endocrine problem)

➢ 19 nortesterones **3.Maternal tumors** (ovarian Gonadoblastoma)	acute regulatory protein (StAR) **3. Anatomic** ➢ 46 XY pure gonadal dysgenesis (appear female) ➢ 46 XY mosaic (45 X/46 XY) or iso-chromosome

INVESTIGATIONS

Confirmatory	Supportive
1)Karyotyping for genotype	**1) RFTs ,BSR, ABGs & electrolytes** for CAH
2)Pelvic ultrasound for internal organs	**2) Hormonal testing for CAH** 17OH progesterone, Cortisol, ACTH, plasma renin activity (PRA), Testosterone, DHT, DHEA, Androstenedione
3)FISH analysis for bar bodies	**3) Provocation tests** HCG stimulation for testosterone or DHT synthesis Ratio of androstenedione to testosterone (elevated in 17-HSD)
	4) Specialised tests Androgen binding studies Androgen receptor gene mutation analysis

TREATMENT

I. MDT (Pediatrician, Surgeon, Endocrinologist, Psychotherapist and Genetician)

II. PARENTAL COUNSELING

Regarding disease, complications, course ,management & **establishing the sex of rearing**

III. TREATMENT OF ACUTE PROBLEMS

Correction of hypoglycemia, dehydration and electrolyte imbalance

IV. SPECIFIC TREATMENT

Medical treatment (Depending upon cause) :
Weekly IM Testosterone injection (PAIS)
Hydrocortisone, Fludrocortisone + sodium supplementation (CAH)
Anti-hypertensives (CAH)

Surgical treatment:
Orchidectomy (PAIS)
Vaginoplasty, Decreasing clitoral size, correction of urogenital sinus (CAH)

TIPS & TRICKS

1) **If Gonads are palpable; Possibilities are XY DSD/Hermaphrodite**.(No palpable gonads; Not male)
2) Must seek permission before exposing genitalia.
3) Generally command for such cases is GPE: Think of DSD if you find nothing on routine GPE especially in case of young child.

STEP IV: RELEVANT GPE

Hands	Hyperpigmentation of knuckles/Axilla (CAH)
Eye	Aniridia, Sunken eyes
CVS	Murmur (in syndromic diagnosis)
Edema	Pedal/sacral

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) Rectal examination (with little finger) to palpate the cervix and confirm a uterus
4) Examine the mother for deep voice, acne and facial hair

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
There is no **distress** or **dysmorphism**.
He is afebrile; **pulse** ___ /min regular in rhythm and normal in volume and character; **B.P** _____ **mm Hg; height** is _____ cm; **weight** is ___kg but I would like to plot them on centile charts.
Inguinal hernia is not present
Stretched phallus length is _____cm
Gonads are/are not palpable in **labio-scrotal folds/inguinal region** (If palpable: They have **pre-pubertal/pubertal volume** bilaterally)
Patient has **fused/bifid labioscrotal folds with/without rugae with single/double opening;** making it **Prader stage** _____
Pubic hairs are present/not present (If present: They are **hypo/hyper pigmented & curled starting from** base of phallus and **spreading to** mid-thigh/umbilicus).
Abdomen is soft, non-tender with no visceromegaly or **palpable mass.**
There is no evidence of **pallor, dehydration, hyper-pigmentation, asymmetry of limbs ,aniridia, murmur** or **edema.**

DIFFERENTIALS

Virilised female	**Testicular failure** (undervirilised males)
1. Congenital adrenal hyperplasia (CAH)—deficiency of: ➢ 21-alpha-hydroxylase ➢ 11-beta-hydroxylase ➢ 3-beta-hydroxysteroid-dehydrogenase **2. Androgen exposure in utero** (maternal androgens): ➢ Progesterone ➢ Medroxyprogesterone	**1. Partial androgen insensitivity** (PAIS) **2. 5-alpha reductase deficiency** **3. CAH**—incomplete deficiency of: • 20,22 desmolase • 3-beta-hydroxysteroid-dehydrogenase • 17-alpha-hydroxylase • 17-beta-hydroxysteroid-dehydrogenase • 17,20 lyase • Lipoid adrenal hyperplasia (deficiency of steroid

AMBIGUOUS GENITALIA (DSD)

1) Inspection
2) Genitalia
3) Abdomen
4) Blood Pressure
5) Relevant GPE

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient [Lying/Sitting on couch/parent's lap]**
d. **Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)**
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: INSPECTION

1) **Face**: Syndromal diagnosis: head, heart, hands (e.g. Beckwith)
2) **Hydration**: Dehydration secondary to vomiting (CAH)
3) **Respiratory rate**

STEP II: GENITALIA (Frog-legged position)

Tell examiner: *'I would like to expose the genitalia for detailed examination' + Seek informed consent of child/parents*: (**Ideally**: Put on gloves while talking)

Inspect	**Prader grading** of virilisation of external genitalia
Inguinal Hernia	*'Chehraa aik side per kar k khansi karain' (Elder child)* *Straighten arms of child above head (To make small child cry; who is unable to obey command)*
Stretched Phallus length	Using a marked tongue depressor
Palpate gonads	Palpate in scrotum and/or in inguinal canal
Urogenital sinus	Gently open cleft/sinus; Locate the urethral meatus Confirm impression of Prader stage (single opening/separate opening) *Skin tags with purplish tinge imply hymen present.*
Pubic hair	Color, shape, spread

Remove gloves

STEP III: ABDOMEN

1) **Skin pinch for dehydration**
2) **Abdominal mass**

STEP IV: BLOOD PRESSURE

Very Important step (Don't miss): Elevated in CAH due to 11 beta-hydroxylase deficiency

Treat CCF (Digoxin, Diuretics, After load reducing medicines e.g. Sildenafil, Captopril)

Large Defects (Surgical management) (at 3–6 months of age)

Prosthetic patch closure (performed at 3months of age, before the pulmonary hypertension causes pulmonary vascular disease (Eisenmenger syndrome)

Muscular defects, can be closed with devices placed at cardiac catheterization

Indications:

General	Failure to thrive
Heart	Failure to control CCF with medicines Supracristal VSD Pulmonary stenosis Aortic regurgitation
Pulmonary HTN	Rising Pulmonary HTN at age of 6-12 months Pulmonary blood flow double the systemic at age >2yr

NOTES

Thrill	**Thrill along left parasternal border**
Heave	No heave (present in Pulmonary HTN)
S1	S1 is soft
S2	**N/Loud P2** if Pulmonary HTN
S3,S4	No added sounds
Murmur	There is a **grade 4/6 pan systolic murmur, best heard in Left lower parasternal area with No specific radiation**
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is not raised
Abdomen	There is no hepatosplenomegaly & Pedal edema
GPE	He is **pale; oral hygiene is good/poor** No evidence of cyanosis, clubbing, splinter haemorrhages, Jane way lesions, petechiae , rash, joint swelling, subcutaneous nodules or any abnormal movement
Gait	Normal

DIFFERENTIALS

VSD
CONGENITAL MR
MR (RHD)
PDA
MVP/DCMP

INVESTIGATIONS

1) **Chest X-ray :** Normal/mild cardiomegaly/cardiomegaly, Enlarged left atrium & ventricle (hypertrophy due to Pul HTN/Increased flow), Increased pulmonary artery silhouette, ↑Pulmonary vascularity (large proximal, small distal arteries)
2) **ECG :** Normal /Biventricular hypertrophy (notched/peaked P waves) by 2 months of age
   Rt ventricular hypertrophy when Pulmonary vascular resistance is high (large defects)
   Left axis deviation in endocardial cushion defects (e.g. Down Syndrome)
3) **Echocardiography :** Position & size of VSD
4) **Doppler echocardiography**: To assess haemodynamic effects
5) **Cardiac catheterization** : To measure O_2 saturation, Pressures in different chambers
6) **Blood CP** (IE)
7) **Blood Cultures** (IE)

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. MEDICAL & SURGICAL THERAPY

Small Defects

Maintaining good dental hygiene (To prevent IE)
Reassure parents Encourage to live normal life
Defect may close spontaneously by 1 year of age
(If don't close: surgical closure may not be required)

Large Defects (Medical management)

Maintenance of Normal growth

VENTRICULAR SEPTAL DEFECT (VSD)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient : **45 degree supine & arms abducted**
d. Exposure: (Ideal exposure in male child: **Shirt off**) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **pink** in room air and there **is/no distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with RR ___ /min, BP ____mmHg with normal Pulse Pressure Pulse rate is ___/min with regular rhythm, normal volume and character. There is no Radio-radial or Radio-femoral delay
Precordium	No precordial bulge, scars marks or pulsations (**Scar=PA banding**)
Apex beat	Visible/palpable in 5th ICS lateral to mid clavicular line and it is **Heaving** in character

PROGNOSIS & COUNSELLING

Cyanotic at birth= Severe disease
Kids Surviving 1st year without treatment = Improve due to systemic-pulmonary collaterals = Seldom survive 2nd decade of life without surgery

After successful total correction
Repair before school-age = Good Survival But sudden death may occur due to dysrhythmias.
Generally asymptomatic + able to lead unrestricted lives
5-20 yr post surgery Follow-up: Marked improvement in symptoms generally maintained
Lower than normal (esp in those with delayed repair/ Transannular outflow tract patch)

1) Exercise capacity
2) Maximal heart rate
3) Cardiac output

Adolescence / adulthood (esp with Transannular outflow tract patch) : RV dilation 2° to severe pulmonary regurgitation **(may need pulmonary valve replacement)**
At adulthood : Careful lifelong follow-up by a specialist in adult congenital heart disease

RECENT ADVANCES

Biventricular pacing	A pacemaker is used to resynchronize the activation of the right and left ventricles) to improve hemodynamics in long ventricular conduction delays

NOTES

	with corrective surgery, any residual pulmonic stenosis or VSD.	
	Associated **asplenia, DiGeorge syndrome**	Antibiotics
	Fatal RSV pneumonia with pulmonary hypertension	Ribavirin, RSV immune globulin
4) Brain abscess (Pt above 2 yr) Less common than CVA CT or MRI confirms the diagnosis	Right-to-left shunting Insidious onset Low-grade fever ± gradual change in behavior Acute onset Headache, nausea, and vomiting Seizures; localized neurologic signs	Antibiotics (keep the infection localized) Surgical drainage (Specific therapy)
5) Thrombo-embolic stroke/CVA (Pt less than 2 yr)	Right-to-left shunting or polycythemia Site: Cerebral veins/dural sinuses & occasionally cerebral arteries	Adequate hydration Phlebotomy (volume replacement with albumin or saline)
6) Gingival disease	Polycythemia, gingivitis, bleeding	Dental hygiene
7) Gout	Polycythemia, diuretic agents	Allopurinol
8) Arthritis	Hypoxic arthropathy	None
9) Clubbing	Hypoxic arthropathy	None
10) Growth failure	Failure to thrive, increased oxygen consumption, decreased nutrient intake	Treat heart failure; correct defect early
11) Psychosocial adjustment	Limited activity, peer pressure; chronic disease, multiple hospitalizations, cardiac surgical techniques	Counseling
12) Pregnancy complications	Poor placental perfusion, poor ability to increase cardiac output	Bed rest

BRAIN ABSCESS MANAGEMENT

MEDICAL	SURGICAL
1) Abscess <2cm 2) Neurologicaly fit child 3) No signs of raised ICP 4) <2 wk duration of illness	1) Abscess >2.5 cm in diameter 2) Multiloculated lesion 3) Located in the posterior fossa 4) Gas is present in the abscess 5) Fungus is identified
Sulbactum –ampicillin alone 3rd generation cephalosporin + Metronidazole (4-6WEEKS) Meropneum +/- vanco to cover MRSA	Encapsulated abscess: Antibiotic+Aspiration *Follow up neuro imaging if not decreasing then surgery*

VII. FOLLOW UP/MONITORING OF DISEASE ACTIVITY

1) Anthropometry
2) To assess RV dilation/dysfunction & PR:
   a. Serial echocardiography
   b. Quantitative magnetic resonance angiography
3) Monitor INR and PT,PTTK if on anti-coagulants

	6) Severe residual gradient across the RVOT (Reoperation indicated) 7) Myocardial infarction (from interruption of an aberrant coronary artery) Conduction disturbances 1) Injury to AV node/Bundle of His (Common : close proximity to the VSD) 2) Permanent complete heart block (rare)(R_x= Permanently implanted pacemaker) 3) Transient complete heart block (rare)(associated with late-onset complete heart block and sudden death) 4) Right bundle-branch block (common) Duration of the QRS interval: Predicts residual hemodynamic derangement and the long-term risk of arrhythmia and sudden death Research R_x : **Biventricular pacing** (a pacemaker is used to resynchronize the activation of the right and left ventricles) to improve hemodynamics in long ventricular conduction delays 5) Premature ventricular beats after repair 24 hr electrocardiographic (Holter) monitoring studies to rule out occult short episodes VT Exercise studies to provoke cardiac arrhythmias not apparent at rest 6) Complex ventricular arrhythmias/Severe residual hemodynamic abnormalities R_x : Prophylactic antiarrhythmic therapy, catheter ablation, or implantation of an implantable defibrillator is warranted *Arrhythmias may improve after hemodynamics restored to more normal level*
Indications for Rerepair	1) Significant residual RVOTO 2) Severe pulmonary insufficiency

VII. TREATMENT OF COMPLICATIONS

(rare if complete repair/palliation in 1st few months)

I. CARDIAC COMPLICATIONS OF CYANOTIC CHD

PROBLEM	ETIOLOGY & NOTES	THERAPY
1) Tet spells	At 6-12 mo of age	Tet spell treatment
2) Heart failure	Young infants with "pink" /acyanotic TOF As the degree of pulmonary obstruction worsens with age, the symptoms of heart failure resolve and eventually the patient experiences cyanosis often by 6-12 mo of age.	Anti-failure therapy

II. EXTRACARDIAC COMPLICATIONS OF CYANOTIC CHD

PROBLEM	ETIOLOGY & NOTES	THERAPY
1) Polycythemia	Persistent hypoxia	Phlebotomy
2) Relative anemia	Nutritional deficiency/ Iron-deficiency anemia, frequently with hemoglobin and hematocrit levels in the normal range (but too low for cyanotic heart disease).	Iron replacement keep Hb:Hct ratio 1:3
3) Infectious disease	**Bacterial endocarditis** Sites: RV infundibulum, Pulmonic, aortic, tricuspid valves (rarely) *Endocarditis may complicate palliative shunts or, in patients*	Antibiotics

complications	2) Diaphragmatic paralysis (Nerve Damage) Recurrent laryngeal nerve 3) Horner syndrome (Nerve Damage) Sympathetic trunk 4) Diminished radial pulse (Artery Damage) 5) Long term arm length discrepancy (Physical Damage) 6) Cardiac failure (Post-Op pulmonary over-circulation) 7) Thrombosis of shunt (Rapid cyanosis → emergent surgery)
Benefit	Cyanosis diminishes
Follow up	Continuous murmur over the lung fields (several days after surgery)
Duration of relief	Variable (As the child grows → more pulmonary blood flow is needed → shunt becomes inadequate → Cyanosis

(BlaLock Water Pott) = *A temporary increase in pulmonary blood flow*

Blalock-Taussig Shunt	Rt Subclavian artery to Pulmonary artery anastomosis
Waterston shunt	Ascending aorta to Rt Pulmonary artery anastomosis
Potts shunt	Descending aorta to Lt Pulmonary artery anastomosis
Central shunt	Shunt from the ascending aorta to the main pulmonary artery

V. VALVE REPLACEMENT (for increasing RV dilation & TR)

Stent valves delivered in cardiac catheterization lab

VI. CORRECTIVE SURGICAL THERAPY (4 months - 2 years)

Procedures	1) Relief of RVOTO by resecting obstructive muscle bundles 2) Patch closure of the VSD 3) Valvotomy for stenotic pulmonary valve 4) Valvectomy if the pulmonary valve annulus is too small OR the valve is extremely thickened (Pulmonary valve annulus split open, and a transannular patch placed across the pulmonary valve ring) 5) MAPCAs ligated
SBE Prophylaxis	**Subacute bacterial endocarditis prophylaxis** is indicated until 6 months after complete repair unless there is a residual VSD. Prophylaxis is then continued as long as there is a residual VSD.
Approach	A right ventriculotomy was once the standard approach A transatrial–transpulmonary approach is routinely performed to reduce the long-term risks of a large right ventriculotomy
Complication of delayed repair	Increased bleeding in the immediate postoperative period (Polycythemia)
Post OP complications Surgical risk In major centers <5%	RV complications 1) Surgically induced PR (pulmonary valvular insufficiency) To-and-fro murmur at LSE due to mild outflow obstruction & mild - moderate pulmonary insufficiency (many pts with TOF repair/all with transannular patch) 2) RV enlargement (2° marked pulmonary valve insufficiency) 3) Tricuspid regurgitation (Tricuspid valve annulus dilates 2° RV enlargement) holosystolic murmur at LLSE 4) RV failure/Heart failure (patients with a large transannular outflow patch) 5) Residual VSD with left-to-right shunting

III. INFANTS WITH TOF

NEONATE WITH SEVERE TOF (urgent medical treatment and surgical intervention)

Aim: Providing immediate increase in pulmonary flow to prevent sequelae of severe hypoxia

1) ABC
2) Prevent Hypothermia (cold➔ increases O_2 consumption➔ additional stress on cyanotic infant)
3) Prevent Hypoglycemia (cyanotic infant is at risk)
4) Prevent Acidosis
5) **IV prostaglandin E1** (0.01-0.20 µg/kg/min) (specific relaxant of ductal smooth muscle➔ dilation of the ductus arteriosus ➔ provides adequate pulmonary blood flow)

Administered intravenously as soon as cyanotic CHD is clinically suspected and continued through the preoperative period and during cardiac catheterization A.E. : Apnea

6) **Surgical intervention**

*Transport to a medical center equipped to evaluate and treat congenital heart diseases

Prolonged, severe hypoxia ➔ Shock & Respiratory failure ➔ Intractable acidosis ➔ Reduced chance of survival

INFANTS WITH SEVERE CYANOSIS IN THE 1ST MO OF LIFE:MARKED RVOTO

Two options are available in these infants.

Critically ill infants/neonates	Infants with less-severe cyanosis
Corrective open heart surgery Benefits: Widespread acceptance Supplanted palliative shunts Excellent short- and long-term results Improved growth of branch pulmonary arteries	Maintain good growth Maintain absence of hypercyanotic spells Elective primary repair at between **4 - 6 mo** of age

INFANTS WITH LESS-SEVERE RVOTO

1) Prevention / treatment of dehydration (to avoid hemoconcentration & thrombotic episodes)
2) Oral propranolol (0.5-1 mg/kg every 6 hr) (to decrease frequency & severity of tet spells)
3) Surgical treatment (Indicated as soon as spells begin)

IV. PALLIATIVE PROCEDURES

Blalock-Taussig shunt	Systemic-to-pulmonary artery shunt performed to augment pulmonary artery blood flow
Rationale of BT shunt	1) To augment pulmonary blood flow 2) To decrease the amount of hypoxia 3) Improve linear growth 4) Augment growth of the branch pulmonary arteries
modified Blalock-Taussig shunt	Most common aortopulmonary shunt procedure and consists of a Gore-Tex conduit/tube anastomosed side to side from the subclavian artery to the homolateral branch of the pulmonary artery/junction? (flow to UL saved?)
Indications of BT shunt	Full repair a higher risk option Patients with comorbidities e.g. other major congenital anomalies or prematurity
Post-OP (BT)	1) Chylothorax (Lymphatic damage)

3. TO RULE OUT COMPLICATIONS

1. **CBC** (Hb ↑, Hct↑, RBCs ↑, TLC N/↑)
2. **BSR** (to rule out hypoglycemia)
3. **Blood C/S** (For I.E.)
4. **Uric acid levels** (to rule out GOUT)
5. **ABGs** (Metabolic acidosis)
6. **CT Scan/MRI Brain** (To R/O Brain abscess/CVA)

TREATMENT

a) Tabulated Overview

MDT	Supportive R_x	Specific R_x	R_x of Complications	F/U?
	1) Tet spells M_x 2) Infants with TOF 3) Palliative procedures 4) Valve replacement	1) Corrective surgery	1) Cardiac 2) Extracardiac	

b) Details (Treatment depends on the severity of the RVOTO)

I. MDT (Multi-disciplinary team approach)

Pediatrician	Paeds cardiologist	Clinical hematologist	Cardiac surgeon
Physical therapist	Dietician	Social worker	Nurse clinician
Psychologist	Psychiatrist		

II. MANAGING TET SPELLS (First 2 yrs, In the morning/after excessive crying)

Admit to ICU (Tet spell : **Calmly Kicks Our National MNA & PM**)

Clothing	Loose any constrictive clothing
Calming	Calm the Patient
Knee-chest	Knee-chest position (↓VR, ↑PVR: decreased shunting)
Oxygen	Corrects relative hypoxemia+ Pulmonary vasodilator
N/saline	Normal saline to treat shock/dehydration (15-20ml/kg)
Morphine	**S/C @ 0.2 mg/kg** (**0.1mg/kg IV** morphine/nalbuphine + **0.1mg/kg IV** metocloperamide) (To relieve anxiety, Prevent sympathetic overdrive, to relax the pulmonary infundibulum)
IV Sodium bicarbonate ($NAHCO_3$)	*Metabolic acidosis develops at arterial PO_2 <40 mm Hg* Rapid correction (within several minutes) if the spell is unusually severe/child shows a lack of response to the foregoing therapy
	Recovery from the spell is usually rapid once the pH has returned to normal.
Propranol	IV Propranolol (**0.1 mg/kg** given slowly to a max of 0.2 mg/kg) (β-blocker) Tab Propranolol (inderal) 10mg @ **1mg/kg** (Decreases infundibular spasm) (↓HR & O_2 requirement)
Phenyl-ephrine	α-adrenergic agonist (phenylephrine) (*Inj Synephrine 10mg*) (Improves RV outflow, decreases Rt-to-Lt shunt, improves the symptoms)
Mech.vent	Intubation and sedation for Resistant spell
Sampling at the end	Baseline blood samples (Premature attempts to obtain blood samples may cause further agitation)
	Repeated blood pH measurements (as rapid recurrence of acidosis may ensue)

The occurrence of a cyanotic spell is an indication to proceed with surgical repair.

Indications
1) Pulmonary atresia
2) MAPCAs (**major aortopulmonary collateral arteries**)
3) Coronary anomalies
4) Multiple VSDs
5) Branch pulmonary stenosis

3. Selective right ventriculography to demonstrate all of the anatomical features. Contrast medium outlines the heavily trabeculated right ventricle. The infundibular stenosis varies in length, width, contour, and distensibility .The pulmonary valve is usually thickened & annulus may be small.

4. Aortography /coronary arteriography (Outlines the course of the coronary arteries)
In 5-10% of patients with the tetralogy of Fallot, coronary artery abnormalities may be present, most commonly an aberrant coronary artery crossing over the right ventricular outflow tract; this artery must not be cut during surgical repair.
Verification of normal coronary arteries is important when considering surgery in young infants who may need a patch across the pulmonary valve annulus. (usually Echo helps to delineate)

5. Cardiac CT Scan (In patients with pulmonary atresia)
To assess the anatomy of the pulmonary arteries and MAPCAs
Cardiac catheterization with injection into each arterial collateral is indicated.
Complete and accurate information regarding the size and peripheral distribution of the main pulmonary arteries and any collateral vessels (MAPCAs) is important when evaluating these children as surgical candidates.

2. SUPPORTIVE

1. CXR (AP view)
Boot Shaped cardiac silhouette: "coeur en sabot" (**POVR of Boot**)
Normal overall heart size
Narrow base, concavity of the left heart border in the area of pulmonary artery
Hypertrophied RV causes the rounded apical shadow to be uptilted so that it is situated higher above the diaphragm than normal and pointing horizontally to the left chest wall
Oligemic lung fields
The hilar areas and lung fields are relatively clear (diminished pulmonary blood flow or the small size of the pulmonary arteries, or both)
Aorta
Large, Rt sided arch (20%) ➔ an indentation of the leftward-positioned air-filled trachea-bronchial shadow in the AP view.

2. ECG
Right axis deviation
Right ventricular hypertrophy
1) Dominant R wave in right precordial chest leads (Rs, R, qR, qRs)
2) RSR' pattern
3) Positive T wave in leads V3R and V1

P wave is tall and peaked= Right atrial enlargement

Thrill	**Systolic thrill at pulmonic area** (if present)
Heave	**Left parasternal heave**
S1	S1 is normal
S2	**Soft**
S3,S4	No added sounds
Murmur	There is a grade **4/6 ejection systolic murmur, best heard in the pulmonic area**
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is not raised
Abdomen	There is no hepatosplenomegaly & Pedal edema
GPE	He is **pale; oral hygiene is good/poor** No evidence of splinter haemorrhages, Jane way lesions, petechiae , rash, or joint swelling.
Gait	Normal

DIFFERENTIALS

TETRALOGY OF FALLOT
TGA + VSD + PS
TA + VSD + PS
DORV + VSD + PS

INVESTIGATIONS

a) Tabulated Overview

I. FOR DIAGNOSIS	II. SUPPORTIVE	III. TO R/O COMPLICATIONS
1) 2D Echo 2) Angiography 3) Right ventriculography 4) Aortography 5) Cardiac CT Scan	1) Chest X ray 2) ECG	1) CBC 2) BSR 3) Blood C/S 4) Uric acid levels 5) ABGs 6) CT Scan/MRI Brain

b) Details

I. FOR DIAGNOSIS

1.Two-dimensional echocardiography (Establishes the diagnosis)
Information about the extent of aortic override of the septum, the location & degree of RVOTO, size of the pulmonary valve annulus and main and proximal branch pulmonary arteries, side of the aortic arch and presence/absence of PDA (supplying a portion of pulmonary blood flow)
In a patient without pulmonary atresia, echocardiography usually obviates (Prevents) the need for catheterization before surgical repair.

2. Cardiac catheterization/Angiography (In patients with pulmonary atresia)
Catheterization is necessary to image the source of blood supply to & size of each lung segment.
RV systolic pressure = Systemic pressure (as RV is connected dire ctly to the overriding aorta)
Pulmonary artery pressure: markedly decreased (5-10 mm Hg)
(Crossing the RVOT, especially in severe cases, may precipitate a tet spell)
Systemic oxygen saturation

Pink tets : Normal	Moderately cyanotic patient at rest : 75-85%

TETRALOGY OF FALLOT (TOF)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient : 45 degree supine & arms abducted**
d. **Exposure:** (Ideal exposure in male child: **Shirt off**) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **Cyanosed & clubbed** but there is no distress or dysmorphism.
Vitals	He is afebrile (to touch) with RR ___ /min, BP ____mmHg with normal Pulse Pressure Pulse rate is ___/min with regular rhythm, normal volume and character. There is no Radio-radial or Radio-femoral delay
Precordium	**Left precordium is bulging** but there are no scars marks ± pulsations
Apex beat	Visible/palpable in 5th ICS lateral to mid clavicular line and it is Normal in character

Thrill	Thrill in left infra-clavicular region
Heave	No heave present in Pulmonary HTN
S1	S1 is normal
S2	**N/Loud in** Pulmonary HTN
S3,S4	No added sounds
Murmur	There is a **grade 4/6 continuous murmur, best heard in left infraclavicular area with radiation to neck and back.**
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is not raised
Abdomen	There is no hepatosplenomegaly & Pedal edema
GPE	He is **pale; oral hygiene is good/poor** No evidence of cyanosis, clubbing, splinter haemorrhages, Janeway lesions, petechiae , rash, joint swelling, subcutaneous nodules or any abnormal movement
Gait	Normal

DIFFERENTIALS

PDA
VSD (subaortic)
AP Window defect
Venous hum

INVESTIGATIONS

1) **Chest x-ray**: Full pulmonary artery silhouette and increased pulmonary vascularity (Moderate to large shunts)
2) **ECG** : Normal to evidence of left ventricular hypertrophy)(right ventricular hypertrophy If pulmonary hypertension is present
3) **Echocardiography :** Position & size of PDA
4) **Doppler echocardiography**: To assess haemodynamic effects
5) **Cardiac catheterization** :To measure O_2 saturation, Pressures in different chambers

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. MEDICAL & SURGICAL TREATM

Medical therapy

Diuretics (Moderate and large PDAs)
Indomethacin/Ibuprofen?

Surgical management

Elective closure of small, hemodynamically insignificant PDAs is controversial.
Cardiac catheter: Coil embolization or a PDA closure device at 1yr (Most PDAs)
Surgical: ligation (rarely)

NOTES

PATENT DUCTUS ARTERIOSUS (PDA)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient : 45 degree supine & arms abducted**
d. **Exposure:** (Ideal exposure in male child: **Shirt off**) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **pink** in room air and there **is/no distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with RR = ___ /min, BP ____mmHg with **normal/wide Pulse Pressure** Pulse rate is ___/min with regular rhythm, **normal/high volume** and of **collapsing character**. There is no Radio-radial or Radio-femoral delay
Precordium	No precordial bulge, scars marks or pulsations
Apex beat	Visible/palpable in 5th ICS lateral to mid clavicular line and it is **Normal/ heaving** in character

Indication to intervene
Clinical signs and hemodynamic evidence of severe obstruction but before severe manifestations

Surgical valvotomy/balloon catheter mitral valvuloplasty
For symptomatic, stenotic, pliable, noncalcified valves of patients without atrial arrhythmias or thrombi

Valve replacement is avoided unless absolutely necessary

RF prophylaxis + IE* prophylaxis (***prosthetic MV replacement**)

NOTES

Thrill	**Diastolic thrill** at apex
Heave	Heaving apical LV impulse
S1	S1 is **Loud**
S2	**N/Loud P2** if Pulmonary HTN
S3,S4	No added sounds
Murmur	There is a **grade 4 mid diastolic murmur with presystolic accentuation, best heard in the mitral area; low pitch rumbling in character, best heard with bell of stethoscope in left lateral position and during expiration**
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is **not raised/raised**
Abdomen	There **is/no hepatosplenomegaly** & Pedal edema
GPE	He is pale; oral hygiene is good/poor No evidence of cyanosis, clubbing, splinter haemorrhages, Jane way lesions, petechiae , rash, joint swelling, subcutaneous nodules or any abnormal movement
Gait	Normal

DIFFERENTIALS

Mitral Stenosis (RHD)
TS (rare)

INVESTIGATIONS

1)CXR
Normal in mild lesion
LA enlargement
Prominence of PA & Rt heart
Greater perfusion of lung apices (reverse of Normal 2^0 obstruction)
Calcifications of the mitral valve

2)Electrocardiogram (ECG)
Normal in mild lesion
Prominent and notched P waves
Signs of RVH
Atrial fibrillation (late)

3) Echocardiography
Thickening of the mitral valve
Narrowing of the mitral orifice during diastole
LA enlargement
Doppler : Transmitral gradient

4)Cardiac catheterization
Quantitates diastolic gradient (MV)
Calculation of valve area
Degree of elevation of pulmonary arterial pressure

TREATMENT

I. MDT (Multi-disciplinary team approach)
II. MEDICAL & SURGICAL TREATMENT

MITRAL STENOSIS (MS)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient : 45 degree supine & arms abducted**
d. **Exposure: (Ideal exposure in male child: Shirt off) [Seek parent's help to undress] (Beware in case of elder girl)**
e. **Right side to approach for examination**

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **pink** in room air and there **is/no distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with RR = ___ /min, BP ____mmHg with normal Pulse Pressure Pulse rate is ___/min with regular rhythm, **normal/Low volume** and character. There is no Radio-radial or Radio-femoral delay
Precordium	No precordial bulge ,scars marks or pulsations
Apex beat	Visible/palpable in 5th ICS lateral to midclavicular line and it is **tapping in character**

Treating infective endocarditis

Surgical treatment

Indications

1) Persistent heart failure
2) Dyspnea with moderate activity
3) Progressive cardiomegaly
4) Pulmonary hypertension

Procedures:

1) **Annuloplasty**
2) **Valve replacement**

RF prophylaxis + IE* prophylaxis (***prosthetic MV replacement)**

NOTES

Thrill	**Systolic thrill** at apex
Heave	Heaving apical LV impulse
S1	S1 is soft
S2	**N/Loud P2** if Pulmonary HTN
S3,S4	No added sounds/ **Prominent S3**
Murmur	There is a **grade 4/6 pan systolic murmur, best heard in the mitral area with radiation towards the axilla and its intensity is increased in left lateral position and with breath held in expiration.**
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is not raised
Abdomen	There is no hepatosplenomegaly & Pedal edema
GPE	He is **pale; oral hygiene is good/poor** No evidence of cyanosis, clubbing, splinter haemorrhages, Jane way lesions, petechiae , rash, joint swelling, subcutaneous nodules or any abnormal movement
Gait	Normal

DIFFERENTIALS

Mitral Regurgitation (RHD)
Congenital MR
VSD to be ruled out

INVESTIGATIONS

1)CXR
Normal in mild lesion
Prominence of the LA and LV
Congestion of peri-hilar vessels (PH) Calcification of the mitral valve(rare)
2)Electrocardiogram (ECG)
Normal in mild lesion
Prominent bifid P waves
Signs of LVH & RVH (If PH)
3)Echocardiography
Enlargement of LA & LV
Abnormally thickened mitral valve,
Doppler studies show severity of MR
4) Heart catheterization & left ventriculography
Considered only if diagnostic questions are not totally resolved by noninvasive assessment.

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. MEDICAL & SURGICAL TREATMENT
Medical Rx (For mild MR)
Prophylaxis against recurrences of RF
Anti-failure medicines
(ACEi & ARBs reduce the regurgitant volume + preserve LV function)
Anti- arrhythmics

MITRAL REGURGITATION (M R)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient : **45 degree supine & arms abducted**
d. Exposure: (Ideal exposure in male child: **Shirt off**) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **pink** in room air and there **is/no distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with RR = ___ /min, BP ____mmHg with normal Pulse Pressure Pulse rate is ___/min with regular rhythm, normal volume and character. There is no Radio-radial or Radio-femoral delay
Precordium	**Left precordium is bulging** but there are no scars marks ± pulsations
Apex beat	Visible/palpable in 6th ICS lateral to midclavicular line and it is **heaving in character (ill-sustained)**

Thrill	No thrill
Heave	**Prominent right ventricular impulse** at LLSE
S1	S1 is normal
S2	**Soft**
S3,S4	No added sounds
Murmur	There is a **Soft Grade 2/6 ejection systolic murmur** (ESM), **best heard in the in ULSE** with no radiation.
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is not raised
Abdomen	There is no hepatosplenomegaly & Pedal edema
GPE	He is **pale; oral hygiene is good/poor** No evidence of cyanosis, clubbing, splinter haemorrhages, Jane way lesions, petechiae , rash, joint swelling, subcutaneous nodules or any abnormal movement
Gait	Normal

DIFFERENTIALS

ASD
Pulmonary Stenosis

INVESTIGATIONS

1) **Chest x-rays** : Cardiomegaly, right atrial enlargement, prominent pulmonary artery, increased pulmonary vascular markings
2) **Electrocardiogram** : Right axis deviation due to right ventricular enlargement
3) **Echocardiogram** : Position & size of ASD
4) **Doppler studies** : To assess haemodynamic effects
5) **Cardiac catheterization** : To measure O_2 saturation, Pressures in different chambers

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. MEDICAL & SURGICAL TREATMENT

Recommendation for closure: Significant shunt around 3 years of age
Secundum ASDs :Closed with an ASD closure device in the catheterization laboratory
Primum & sinus venosus defects: Surgical closure

NOTES

ATRIAL SEPTAL DEFECT (ASD)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient : **45 degree supine & arms abducted**
d. Exposure: (Ideal exposure in male child: **Shirt off**) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **pink** in room air and there **is/no distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with RR = ___ /min, BP ____mmHg with normal Pulse Pressure Pulse rate is ___/min with regular rhythm, normal volume and character. There is no Radio-radial or Radio-femoral delay
Precordium	No precordial bulge, scars marks or pulsations
Apex beat	Visible/palpable in 5th ICS lateral to mid clavicular line and it is Normal in character

Thrill	**Systolic thrill** at aortic area radiating to neck
Heave	There is no left parasternal heave *(present in Pulmonary Hypertension)*
S1	S1 is normal
S2	**A2 is soft**
S3,S4	No added sounds
Murmur	There is a **grade 4/6 Ejection Systolic murmur, best heard in the aortic area/ LUSE** with **radiation towards the carotids** and **its intensity is increased in expiration & on leaning forward.** There is also **ejection systolic click** at aortic and apical area
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is not raised
Abdomen	There is no hepatosplenomegaly & Pedal edema
GPE	He is **pale; oral hygiene is good/poor** No evidence of cyanosis, clubbing, splinter haemorrhages, Jane way lesions, petechiae , rash, joint swelling, subcutaneous nodules or any abnormal movement
Gait	Normal

DIFFERENTIALS

Aortic Stenosis (RHD)
Co-arctation of Aorta
Hypertrophied Obstructive CardioMyopathy

INVESTIGATIONS

1) **Chest x-rays** Normal Heart size, Prominent Ascending aorta
2) **Electrocardiogram** LVH
3) **Echocardiogram** LVH, AS
4) **Doppler studies** For Degree of aortic stenosis
5) **Cardiac catheterization** when the echo data are equivocal

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. MEDICAL & SURGICAL TREATMENT

1) Balloon valvuloplasty
2) Aortic valve replacement
3) Ross procedure (Diseased aortic valve is replaced with the person's own pulmonary valve)

NOTES

AORTIC STENOSIS (AS)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient : 45 degree supine & arms abducted**
d. **Exposure: (Ideal exposure in male child: Shirt off)** [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **pink** in room air and there **is/no distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with RR = ___ /min, BP ____mmHg with **Narrow Pulse Pressure** Pulse rate is ___/min with regular rhythm, normal volume and **slow rising** in character. There is no Radio-radial or Radio-femoral delay
Precordium	No precordial bulge, scars marks or pulsations
Apex beat	Visible/palpable in 5th ICS lateral to midclavicular line and it is **heaving in character (Well-sustained)**

TIPS & TRICKS

1) AR is picked by peripheral signs of high volume bounding pulses.
2) Clinical features include history of angina or strain.

NOTES

Thrill	**Diastolic thrill** at aortic area (?) *(Say if thrill is felt)*
Heave	There is no left parasternal heave *(present in Pulmonary Hypertension)*
S1	S1 is normal
S2	**A2 is soft**
S3,S4	No added sounds
Murmur	There is a **grade 4 early diastolic murmur, best heard in the aortic area/ LUSE** and **its intensity is increased in expiration & on leaning forward. Pistol shot sounds** are heard over femoral artery.
Back	Bases of lungs are clear and there is no sacral edema
JVP	JVP is not raised
Abdomen	There is no hepatosplenomegaly & Pedal edema
GPE	He is **pale; oral hygiene is good/poor** No evidence of cyanosis, clubbing, splinter haemorrhages, Jane way lesions, petechiae , rash, joint swelling, subcutaneous nodules or any abnormal movement
Gait	Normal

DIFFERENTIALS

Aortic Regurgitation (RHD)
Pulmonary Regurgitation needs to be ruled out (RVH,P2 loud, murmur in Pulmonary area)

INVESTIGATIONS

1) **Chest x-rays** Enlargement of LV and aorta
2) **Electrocardiogram** Normal/LV hypertrophy and strain with prominent P waves
3) **Echocardiogram** Large LV and diastolic mitral valve flutter or oscillation caused by regurgitant flow hitting the valve leaflets.
4) **Doppler studies** For Degree of aortic runoff into the left ventricle
5) **Magnetic resonance angiography** To quantify regurgitant volume
6) **Cardiac catheterization** when the echo data are equivocal

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. MEDICAL & SURGICAL TREATMENT

Mild and moderate lesions (Well tolerated)
Afterload reducers (ACEi,ARBs)
Prophylaxis against recurrence of acute rheumatic fever
Surgical intervention /Valve replacement
Indications
1) Advanced heart failure, pulmonary edema, or angina
2) ST-T wave changes on ECG
3) Signs of ↓ myocardial performance (↑ LV dimensions on echo, ↓EF)

PROGNOSIS

Unlike mitral insufficiency, aortic insufficiency does not regress.
Patients with combined lesions during ARF may have only aortic involvement 1-2 yr later.

AORTIC REGURGITATION (AR)

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient : 45 degree supine & arms abducted**
d. **Exposure:** (Ideal exposure in male child: **Shirt off**) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

See Cardiovascular system exam for details

Redress the child and say thank you!

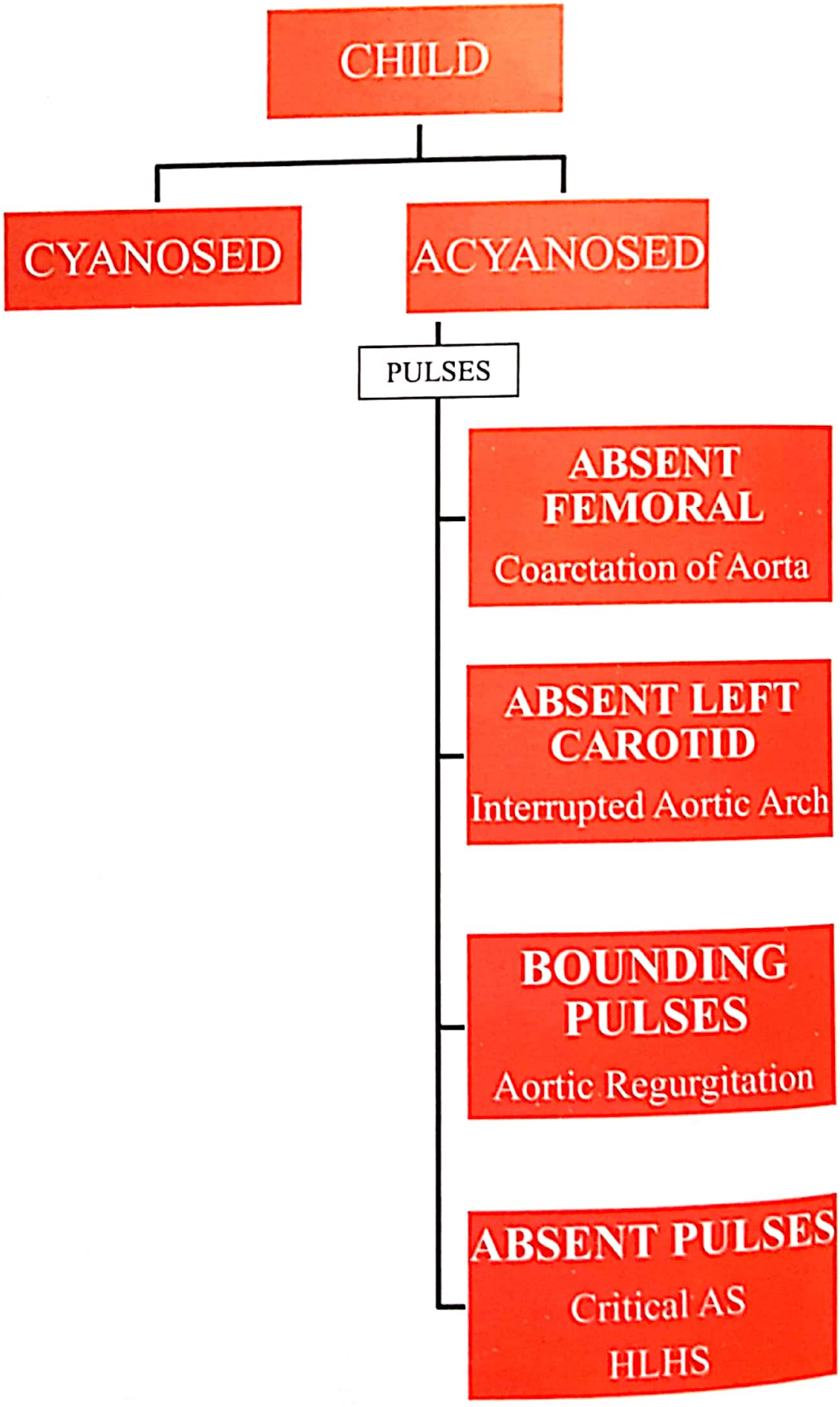
OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [For Haematuria; Glucose; Protein]
5) **Motor system examination** (If any focal deficit)

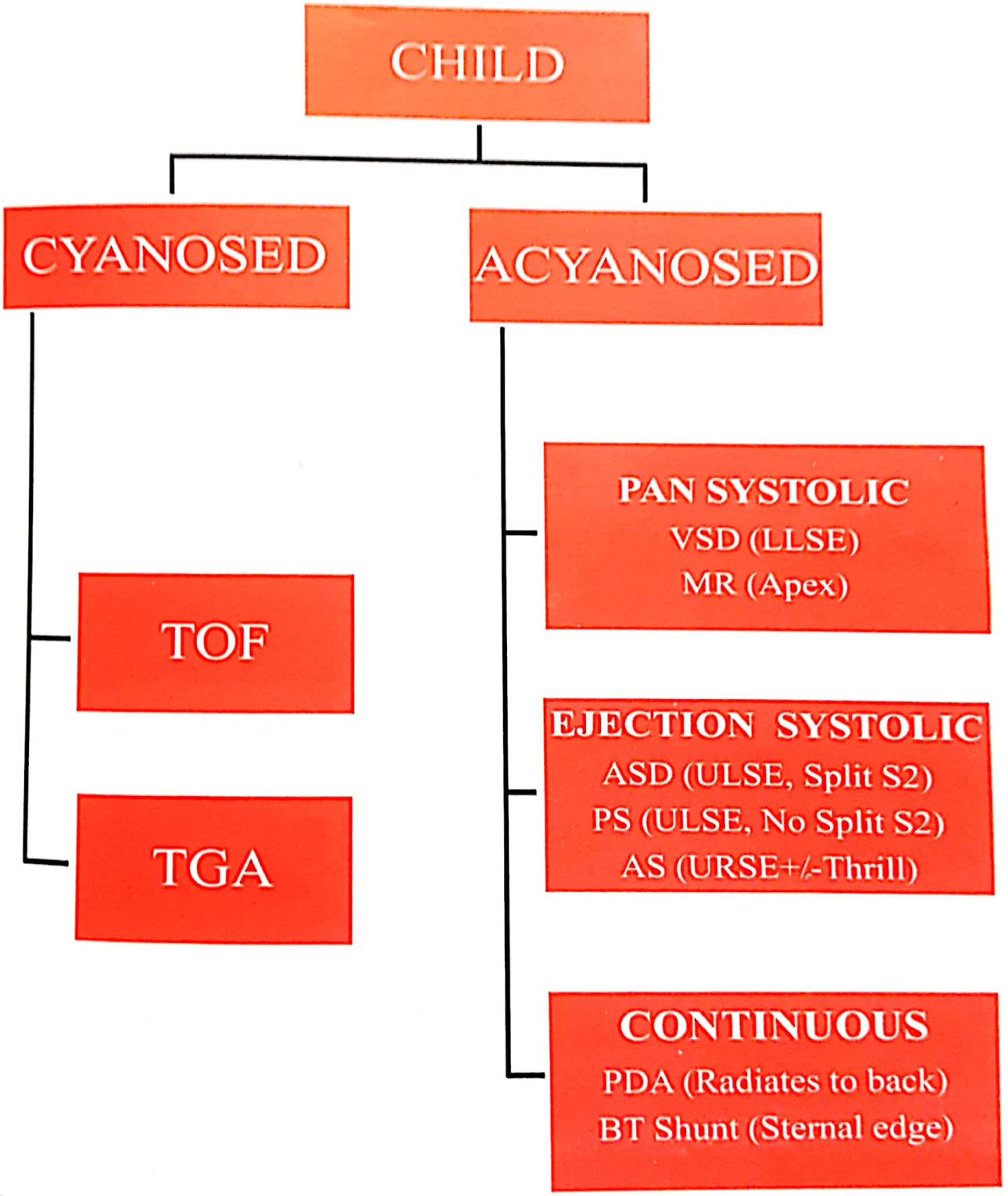
DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)

General look	I have examined _____, __ years old who is **conscious, cooperative,** having normal/thin **built** and a **cannula** in his _____. He is **pink** in room air and there **is/no distress** or **dysmorphism.**
Vitals	He is afebrile (to touch) with RR = ____ /min, BP ____mmHg with **Wide Pulse Pressure** (↑ Systolic blood pressure, ↓ diastolic pressure) Pulse rate is ___/min with regular rhythm, **high volume** and **Collapsing** character. There is no Radio-radial or Radio-femoral delay
Precordium	**Left precordium is bulging** but there are no scars marks ± pulsations
Apex beat	Visible/palpable in 6th ICS lateral to midclavicular line and it is **heaving in character (ill-sustained)**



APPROACH TO CVS EXAM



lightly)	
Benign Murmur	**Pathological Murmur**
Systolic S1 S2 : Normal Soft <3/6 in intensity Otherwise healthy Often position dependant	Diastolic Abnormal S1 + S2 Loud >3/6 Abnormal cardiac exam

ABDOMEN

Hepatomegaly (gives idea of Right sided heart function)

Normal

Newborn <2cm below Right SCM	Child: Not palpable

Liver span (Percuss)

Newborn	4-5 cm
12 year old	6-8 cm (girls uptil 7cm)

Splenic enlargement= Palpable tip >2cm below Left SCM

HANDS AND SKIN

Clubbing	Lovibond's angle (N=160 degree, Clubbing >180 degree)
Splinter haemorrhages	Linear, reddish brown marks along the axis of the finger and toenails, thought to be due to circulating immune complexes (IE, vasculitic disorders, Trauma)
Jane way lesions	Painless red spots, which blanch on pressure, on the thenar/ hypothenar eminences of the palms, and soles of the feet. (IE)
Osler's nodes	Painful raised erythematous lesions which are rare but found most often on the pads of the fingers and toes.(IE)
Xanthomata	Yellow skin or tendon nodules from lipid deposits at the palmar and extensor surfaces of the hands
Fever	Infective endocarditis , Pericarditis
Warm & sweaty	Autonomic stimulation
Cold & clammy	Hypotension and shock
Petechial rash	Legs and conjunctivae (vasculitis, IE)

THE FACE AND EYES

Corneal arcus	A creamy yellow discoloration at the boundary of the iris and cornea caused by cholesterol deposition. It is more common in men and black
Xanthelasmata	Soft yellowish plaques periorbitally and on the medial aspect of the eyelids associated with hyperlipidaemia *If present check the patellar & Achilles tendons for xanthomata*
Central cyanosis	Congenital heart disease, heart failure
Roth's spots	Flame-shaped retinal haemorrhages with a 'cotton-wool' centre (IE, Anemia or Leukaemia)

ARTERIAL PULSES

Pulse	A pressure wave that can be felt due to ejection of blood from the left ventricle into the systemic arterial circulation **Children's pulse can vary significantly with respiratio n** **Inspiration**: (*Inspires*) Rate speeds up **Expiration**: Rate slows

1) **severity of that lesion.** For example, with a ventricular septal defect, assess thesize of the shunt,; with pulmonary stenosis, assess the severity by the timing of the peak of the murmur, the associated presence or absence of a click, movement of S2 with respiration, and clinical signs of right-ventricular hypertrophy.
2) **Doubt about pulse**: Palpate your own pulse at the same time. If it is not synchronous with yours, it is the patient's.
3) **Measuring reliable heart rate:** In children radial pulses are sometimes not a good source of knowing heart rate especially during heat of exam so during auscultation measure heart rate from apex but do feel the pulse to complete exam.
4) **Grading systolic murmur clinically**: *'Look for Thrill to grade it'*

No Thrill (Grade 1-3)		**Thrill present** (Grade 4-6)	
Grade 1	Faint, picked by experts	**Grade 4**	Very loud with thrill
Grade 2	Faint, but is immediately audible	**Grade 5**	Audible with edge of stethoscope
Grade 3	Moderately loud: No thrill	**Grade 6**	Audible with stethoscope just removed from contact with chest

VIVA QUESTIONS

ANTHROPOMETRY IN CVS	
Height	**Short** (Down, Noonan, Turner; CHD causing FTT, e.g. large atrioventricular canal)
	Tall (Marfan; check arm span if you suspect this)
Weight	**Failure to thrive** (congenital rubella, severe heart disease, cyanosis or CCF)
FOC	**Small** (congenital rubella)
INSPECTION	
Dysmorphic	Down, Noonan, Marfan, Turner, Williams, Alagille, NF -1
Scars	• **Median sternotomy** (all open heart corrections) • **Lateral thoracotomy** (e.g. coarctation repair, pulmonary artery banding, ligation of PDA or vascular ring, pulmonary artery reconstruction, shunts) • **Groin** (cardiac catheters)
PRECORDIUM	
Apex beat	Lowermost & outermost part of precordium where a definitive impulse is felt. *(Deviation of apex beat=Rt/Lt/generalized ventricular enlargement)*
	Apex beat : In children >7yr Located in 5th ICS in mid-clavicular line Diameter <2cm Tapping quality Duration <2/3rd of systole
S1	AV valve closure before ventricular systole
S2	Aortic/Pulmonary valve closure at ventricular diastole
S3	Volume overloaded non-contracting ventricles: causing AV valvular leak
S4	Atrial contraction against hypertrophied left ventricle (HTN) ; occurs before S1
MURMURS	
Diaphragm	For **High** pitched sounds e.g. pericardial rubs, S1,S2, Most murmurs
Bell (placed	For **Low** pitched sounds e.g. gallop, S2 split, S2 wider split end of inspiration

1) **Is the Child in failure?** Very favourite exam question. Be prepared for it. Tachypnea, Tachycardia and Hepatomegaly are three signs of failure.
2) **Locating Apex Beat**: In almost all cases in exams, apex beat is visible so don't waste your time; Inspect and place your finger on the apex beat but keep in mind that it should be lower and outermost point.
3) **Thrill**: Thrill in 2nd Left ICS is palpable P2; signifying severe pulmonary HTN.
4) **Best heard Areas**: First heart sound is best heard at apex and second heart sound at pulmonary area.
5) **Loud P2**: In case of loud P2, always auscultate in pulmonary and aortic areas. If 2nd heart sound appears loud in pulmonary area, compared to aortic area, say loud second heart sound.
6) **Murmur heard on placing stethoscope:** on precordium is usually systolic murmur.
7) **Never forget to palpate carotid during auscultation** (Make it a habit). If you cannot palpate carotids or child is too young, palpate brachial pulse.
8) **Checking Back**: When u check radiation of murmur on the back, press at sacrum at the same time, to check for sacral edema to save your time.
9) **Correct coinage**: Do not use abbreviations.
10) **Confused between MR and VSD**: Always check radiation to the axilla in left lateral position.
11) **You do not have to give a specific diagnosis immediately !** e.g. if you are sure that a patient has valvular aortic stenosis, you should say so, but if there is any uncertainty, it is prudent to give as a diagnosis 'left ventricular outflow tract obstruction' (LVOTO) and then proceed to delineate which of the various causes of LVOTO (supravalvular, valvular or subvalvular) is most likely and why.

Supravalvular LVOTO has a thrill	Valvular LVOTO has a click and a thrill	Subvalvular LVOTO has neither

12) **Murmur in the region of the pulmonary valve**: Possibilities include a pulmonary flow murmur, an atrial septal defect (which is technically also a pulmonary flow murmur) or a right-ventricular outflow tract obstructive lesion; it is prudent to describe it initially as a **'right-ventricular outflow tract' (RVOT) murmur** .

Supravalvular : thrill at the upper left sternal border, or in the suprasternal notch	Valvular: systolic ejection click ± thrill	Subvalvular

13) **Cyanosed child with confusing array of murmurs**: You do not have to give an anatomically correct diagnosis. It is better to start in general terms, such as: 'This child has **complex cyanotic congenital heart disease'**. If you have a reasonable idea of the likely anatomical diagnosis, say so, but if not, it is sensible to take each murmur in turn and give a brief differential diagnosis of each (provided that these are relevant to a child with cyanotic congenital heart disease)
14) **The Valsalva manoeuvre** usefulness

Hypertrophic cardiomyopathy (HCM)	Increases intensity of the murmur (increased intrathoracic pressure, decreased venous return & decreased intracardiac volume & more severe LVOTO)
Mitral valve prolapse	Murmur is increased + systolic click is heard earlier
Innocent systolic outflow tract murmurs	Decrease in intensity

With any findings strongly suggestive of a specific diagnosis, make a point of going beyond simple diagnosis of the said lesion, and be aware of clinical signs indicating the

6.BLOOD PRESSURE

Apply age appropriate cuff; inflate 30mmHg more than loss of palpation on palpatory method; apply stethoscope and gradually decrease cuff pressure

STEP V: RELEVANT GPE

BCG scar	
Eye	For xanthelasmata & corneal arcus (at iris) Conjunctiva for pallor , petechiae & jaundice (haemolysis associated with artificial valves)
Oral cavity	Dental hygiene (SBE), Central cyanosis*(ask to turn tongue upward)* If uncertain about cyanosis, comment on the need to look again in natural daylight, if the room is artificially lit. Avoid saying ' pink' or 'blue': say 'not cyanosed' or 'cyanosed'
JVP	**Jugular venous pressure** (JVP) usually performed after 8 yrs of age Sit child at 45° in the standard manner; use torch to illuminate Hepato-jugular reflux ± (Can be skipped)

STEP VI: ABDOMEN

ASK '*Beta pait main kahin daard tuu nahi'*

1) **Hepatomegaly** (congestive cardiac failure) & **span**
2) **Pulsatile liver** (tricuspid incompetence)
3) **Splenomegaly** (SBE)

STEP VII: GAIT

Ask: *'Bachaa Chal saktaa hai?*

Rapid scan to rule out hemiplegia

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) **Fundoscopy** (Features of hypertension, diabetes and Roth's spots)
4) **Urinalysis** [Haematuria (endocarditis, vasculitis); Glucose (diabetes); Protein (hypertension and renal disease)]
5) **Motor system examination** (If any focal deficit)

TIPS & TRICKS

1) **An infant or fractious toddler:** Approach is different: Distant observation is very important, noting size, colour, respiratory rate and perfusion. It is appropriate to tell the examiners that you are going to start with auscultation while the baby is quiet; if the baby does become restless, a breast or bottle may be a life saver.
2) **With a very uncooperative child:** Key is to do what you can, while you can, without becoming angry or overtly frustrated.
3) **When command is pulses**: Think of PDA, CoA and AR.
4) **Very first thing to ascertain on inspection:** *Is the child cyanosed or Not?*

STEP IV: PULSES & BP (In Lying position)

1.NAILS

1) Clubbing & cyanosis (Place hands on own hand)
2) Koilonychias, splinter hemorrhages
3) Capillary refill time

2.HAND

1) Feel Temperature
2) **Dorsal aspect:** xanthomata & petechiae
3) **Palmar aspect of hands and fingers** : Janeway lesions & Osler's nodes
4) **Assess thumbs & radii** (TAR = TOF, ASD)

3.UPPER ARTERIAL PULSES

1) Radial pulse
Slightly flex & pronate hand;locate with your index and middle fingers
Count **R**ate, assess **R**hythm,**V**olume and **C**haracter. ***(Mnemonic: RVChamp)***
BT shunt=absent radial pulse on that side

2) Check for **collapsing pulse** (e.g. AR, PDA) *'Baazuu ya kandhaay main dardd tuu nahi hai?'*
Feel the pulse with the base of your fingers, then raise the patient's arm vertically above the patient's head

3)Check for **Radio-radial delay**

4) Check for **Radio-femoral delay**
'Main aap ki tang wali nabz mehsoos karoon ga'
Approach from head end; Place your index and middle fingers over the femoral artery, which is just inferior to the midpoint between the anterior superior iliac spine and the pubis
Difference in volume (CoA)
Check groin area for cardiac catheters

5) Check both **Brachial pulses**
Flex patient's arm ; use index and middle fingers to palpate this over the lower end of the humerus just above the elbow joint (use opposite hand)

6)Carotid pulse (**one at a time, semirecumbent position)
'Main aap ki gardaan wali nabz check karoon ga, iss say aapko koi takleef nahi ho gi'
At the angle of the jaw, anterior to the sternocleidomastoid muscle.

4.LOWER ARTERIAL PULSES

*(**Tip**: If time is less and command is Not 'Pulses only'; Just feel Dorsalis pedis)*

1) Popliteal pulse
Flex knee at 120 degree and push fingers of both hands into popliteal fossa
2) Posterior tibial
Located 2 cm below and posterior to the medial malleolus
3) Dorsalis pedis
In proximal part of first intermetatarsal space

5.FEET

Edema, Temperature, Clubbing

3. Locate apex beat (measure distance from Lt sternal border & count ribs)
Assess character of apex beat

Normal	
Tapping (Touches)	MS
Heaving (Ill sustained)	MR, AR
Heaving (Well sustained)	AS, HTN

If difficult to ascertain....ask kid to roll over to left side and breathe out

4. Thrill (a palpable murmur caused by turbulent flow)
Best palpated through bone

By thenar area in older child	By tips of finger in younger children

Suprasternal and supraclavicular regions, pulmonary area (closure of the pulmonary valve)

5. Heave (forceful lift associated with dilatation of a heart chamber)
Ulnar border & at eyes at level
Parasternal border and Substernal region

STEP III: AUSCULTATION (PRECORDIUM)

1. LYING POSITION

Warm the diaphragm of stethoscope with clothes & check on dorsal surface of hand.
Must time auscultation with carotids *'Main aap k galaay ki nabz mehsoos karon ga jis say apko koi takleef nahi ho gi'*

Use Diaphragm (Note S1,S2, split ,added sounds, murmurs)
Apex (*Recording heart rate for six seconds and multiplying by 10 at this point is helpful*)
Axilla (Radiation of MR)
Parasternal border
Pulmonary area *('gheray saans laitay rahain')* to check for normal S2 split with respiration?
Left Infraclavicular region (PDA)
Aortic areas
Listen for bruits over both carotid arteries, using the diaphragm **(held inspiration)** *(Lumba sans lain aur rok lain)... Sans lay lain* (V.V.Important)

If murmur is found note point of maximal intensity, grade, radiation (Axillae/carotids)

Use bell
Apex :keeping patientt in left lateral position (MS=mid diastolic rumbling sound ± opening snap)
All areas described above

2. SIT THE CHILD UP

Listen to any murmur's variation with the change in position
(*Ghehra sans lain, bahar nikalain aur rok lain*: Held expiration) = **Place diaphragm at Aortic area & LLSE** (for early diastolic murmur of AR)...*Sans lay lain* (V.V.Important)

3. BACK (with Bell)

Interscapular murmur (Co-arctation of aorta)
Lung Bases with the Bell (for crackles of pulmonary edema) '*Ghehraay sans laitay rahain'*
Sacral edema (using two fingers press gently for few seconds)

CARDIOVASCULAR SYSTEM

1) Inspection
2) Palpation
3) Auscultation
4) Pulses & BP
5) Relevant GPE
6) Abdomen
7) Gait

LISTEN TO COMMAND	
Command: Do CVS	Pulses first
Command: Do precordium/chest	Precordium first

MOST IMPORTANT FIRST THING TO LOOK FOR : **IS THE CHILD CYANOSED???**

Cyanosed : Think of Cyanotic CHD	**Not Cyanosed**: Think of Acyanotic CHD

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient : <u>45 degree supine & arms abducted</u>**
d. **Exposure:** (Ideal exposure in male child: **Shirt off**) [Seek parent's help to undress] (<u>Beware in case of elder girl</u>)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: INSPECTION

<u>From foot end</u>

Dysmorphism; ill/healthy looking; Obese/Thin or wasted; average/tall/short
Respiratory rate (use wrist watch), type of respiration
Symmetry (*Left Precordial bulge = RVH*) (*Right chest prominence: dextrocardia with chronic ventricular hypertrophy*)
Skeletal deformity of chest (*pectus carinatum/excavatum*)

<u>Inspection from side</u>

(at level of patient for Precordium & both axillae)

Scars (BT shunt, Infrascapular (back) = CoA or PDA repair)
Skin
Visible pulsations *(Thin/higher output states e.g. fever)*

STEP II: PALPATION (PRECORDIUM)

Warm hands + Ask: *Beta aap ko chaati main kahin dard tuu nahi?*
1. Trachea: *Main aap ki sans ki nali mehsoos karoon ga!* Check trachea (3 finger method)
2. Dextrocardia check (place both hands on Rt & Lt side of chest)

TREATMENT

I. MDT: Pediatrician,Pediatric neurophysician,Opthalmologist, Pulmonologist, cardiologist

II. SPECIFIC TREATMENT

Everolimus

1) Slows the growth or even reduces the size of SEGAs
2) Effective in treating refractory seizures
3) Reduces the volume of renal angiomyolipomas
4) Reduces the volume of lymphangioleiomyomatosis
5) Reduces the volume of facial angiofibromas

SEGAs

Presymptomatic treatment	Everolimus (effective in slowing the growth or even reducing the size of SEGAs)
Acutely symptomatic SEGA	Surgical resection
Growing but otherwise asymptomatic SEGA	Surgical resection **OR** medical treatment with an mTOR inhibitor

Infantile spasms

Vigabatrin (first-line therapy)
ACTH (if treatment with vigabatrin fails)

Seizures

Anticonvulsant therapy of other seizure types in TSC
Epilepsy surgery (for medically refractory TSC)

Renal angiomyolipoma

Angiomyolipoma with acute hemorrhage	Embolization followed by corticosteroids Nephrectomy should be avoided.
Asymptomatic, growing angiomyolipomas measuring larger than 3 cm in diameter	mTOR inhibitor, Everolimus Selective embolization (alternative) Kidney-sparing resection (alternative)

TIPS & TRICKS

1) If command is motor system then do motor system first then relevant GPE.

NOTES

III. PRENATAL TESTING

1) **Genetic testing** if known TSC mutation exists in that family.
2) **Fetal Echo for rhabdomyoma**

IV.TO RULE OUT COMPLICATIONS

1) **2D-Echo** for **Cardiac rhabdomyomas**
2) **Chest X ray/CT scan** for Pulmonary Lymphangioleiomyomatosis
3) **USG Abdomen/CT scan** for **Renal angiomyolipomas**
4) **Urine RE** for hematuria

DIAGNOSTIC CRITERIA*

'8 Tubers Cafe': Definite diagnosis:

Organ system	Major Features of TSC	Minor Features of TSC
Brain	1)**Cor**tical tuber 2)**Sub**ependymal **N**odule 3)Subependymal giant cell **Astrocy**toma	1)Cerebral **white matter migration lines**
Skin lesions**	4) Sha**Green** patch 5) Hypomelanotic macules (>3)/**Ash leaf** 6) **Facial** angiofibroma or **forehead** plaque 5) **Ungual**/periungual fibroma (non-traumatic)	2)Multiple **dental pits** 3)**Gingival** fibromas 4)**Confetti** skin lesions
Eye	8)Multiple **ReTinal Hamartomas**	5)**Retinal achromatic patch**
Heart	9) Cardiac **Rhabd**omyoma	
Lung	10)**Pulmonary** lymphangioleiomyomatosis	
GIT		6)**Hamartoma**tous rectal polyps
Kidney	11)Renal angiomyolipoma(**AML**)	7)Nonrenal **hamartomas** 8)Multiple renal **cysts**
Bone		9)Bone **cysts**

SKIN LESIONS**

A shagreen patch	Characteristic of TSC consisting of roughened, raised lesion with orange-peel consistency located primarily in lumbosacral region
Hypomelanotic macules/ ash leaf macules (>90% pts)	Hypomelanotic macules likened to an ash leaf on the trunk and extremities. Visualization is enhanced by the use of a Wood ultraviolet lamp. To count as a major feature, at least three hypomelanotic macules must be present.
Facial angiofibromas	Develop between **4 and 6 yr of age**; they appear as tiny red nodules over nose & cheeks and are sometimes confused with acne. Later, they enlarge, coalesce, & assume a fleshy appearance.
Periungal Fibromas (15-20% pts)	During **adolescence or later**, small fibromas or nodules of skin may form around fingernails or toenails in 15-20% of the TSC patients

STEP V: MOTOR SYSTEM

See Motor system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry & progressive centile charts
3) Fundoscopy (For retinal lesions: Hamartomas, White depigmented patches)
4) Developmental assessment
5) Urine & stool exam for blood

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
He is **moving all his limbs** and **interacting appropriately** for his age.
A **5 x 7 cm irregular patch** of _____**color** is present **on Right side of trunk.** (Describe all neurocutaneous lesions found)
Gait and examination of **spine** are normal.
His **FOC** is _____cm, **Height** is ____cm and **Weight** is ____kg. I would like to plot them all on centile charts.
His **RR** is ___/min, **HR** is ___/min with regular pulses; normal in volume and character. **BP** is ____mmHg. I would like to plot it on centile chart.
Eye movements and **visual acuity** are normal.
He has/doesn't have **Periungal Fibromas, dental pits, Gingival fibromas, Murmur, basal crepts, abdominal mass** and/or **edema**.
Motor system examination is unremarkable.

DIFFERENTIALS

Make differentials according to predominant findings:

1) Hypomelanosis of Ito
2) Sturge-Weber syndrome
3) Epidermal nevus syndromes
4) Multiple endocrine neoplasia
5) Isolated brain tumors
6) Cardiac myxoma

INVESTIGATIONS

I. FOR DIAGNOSIS

1) **Clinical Criteria*** (as below)
2) **Brain MRI** confirms the diagnosis in most cases
   It is best way of identifying cortical tubers, which can form before birth.
3) **Genetic testing for TSC1 and TSC2 mutations** (if clinical criteria not met)

II. SUPPORTIVE

Wood ultraviolet lamp for enhanced Visualization of the hypomelanotic macule

TUBEROUS SCLEROSIS

1) General Look
2) Gait + Back
3) Anthropometry
4) Relevant GPE
5) CVS, Chest, abdomen
6) Motor system

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For slurring/poor articulation/intellectual impairment)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **Position patient** [Lying/Sitting on couch/parent's lap]
d. **Exposure:** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Neurocutaneous stigmata (location, size, color), Posture, Autistic behavior, ADHD
Respiratory rate: ? in Pulmonary Lymphangioleiomyomatosis/CCF

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

STEP III. ANTHROPOMETRY

FOC (? in SEN, SEGA, Hydrocephalus)

STEP IV: RELEVANT GPE

Hands	Periungal Fibromas
Pulse rate	? due to Cardiac rhabdomyomas/CCF
B.P.	? due to Renal angiomyolipomas
Eye	Visual acuity, Extraocular muscle movements
Hearing	As part of developmental assessment
Oral cavity	Multiple dental pits, Gingival fibromas
Skeletal system	For Bone cysts
Edema	Due to CCF

STEP V: CVS, CHEST, ABDOMEN

1) **CVS**: Murmur/rub due to Cardiac rhabdomyomas/CCF
2) **CHEST**: Wheeze/Crepts due to Pulmonary Lymphangioleiomyomatosis/CCF
3) **ABDOMEN**: Renal Mass

3) Fundoscopy (Tomato ketchup fundus)
4) IOP measurement
5) Urine & stool exam for blood

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
A **5 x 7 cm irregular patch** of **purple color** is present **on Right side of face** covering forehead, eyelids and cheeks. He is **moving all his limbs**.
Gait and examination of **spine** are normal.
His **FOC** is _____cm, **Height** is ____cm and **Weight** is ____kg. I would like to plot them all on centile charts.
His **RR** is ___/min, **HR** is ___/min with regular pulses; normal in volume and character.There is **no Radiofemoral delay. BP** is ____mmHg. I would like to plot it on centile chart.
Eye movements and **visual acuity** are normal.
Motor system examination is unremarkable.

DIFFERENTIALS

Klippel-Trénaunay-Weber's syndrome (port wine stains, Solid visceral tumors)
Beckwith-Wiedemann syndrome (port wine stain, visceromegaly, hypoglycemia)
Dyke-Davidoff-Masson syndrome (atrophy of a hemispere during infancy)

INVESTIGATIONS

I. FOR DIAGNOSIS
1) **MRI brain with contrast** (imaging modality of choice)
For demonstrating the leptomeningealangioma,White matter abnormalities and atrophy
2) **CT Scan head for calcifications**

II. TO RULE OUT COMPLICATIONS
1) **Ophthalmologic evaluation** for glaucoma (IOP)

TREATMENT

I. MDT: Pediatrician,Pediatric neurophysician,Opthalmologist

II. SPECIFIC TREATMENT

Well controlled seizures + Normal or near-normal development
1) Anticonvulsants
2) Surveillance for complications (glaucoma, buphthalmos, and behavioral abnormalities)
3) Pulsed-dye laser therapy (for facial port-wine stain)

Seizures refractory to anticonvulsant therapy
1) **Hemispherectomy** (especially in 1st 1-2 yr : seizures arise from primarily 1 hemisphere)
2) Regular measurement of intraocular pressure

Pulsed-dye laser therapy (for facial port-wine stain :excellent clearing if located on the forehead)

TIPS & TRICKS

1) If command is motor system then do motor system first then relevant GPE.

STURGE WEBER SYNDROME

1) General Look
2) Gait + Back
3) Anthropometry
4) Relevant GPE
5) Motor system
6) Developmental assessment

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For slurring/poor articulation/intellectual impairment)
   2) *Kis class main parhtay hain?* (For Mental age)
c. Position patient [Lying/Sitting on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Port-wine stain (location, size, color), Posture

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

STEP III. ANTHROPOMETRY

STEP IV: RELEVANT GPE

Hands	Pallor
Pulse	Check peripheral pulses for Coarctation of Aorta
Eye	Visual acuity, Extraocular muscle movements, Corneal clouding, Iris
Oral cavity	Mucosa of mouth & pharynx(capillary malformation)
Trunk	Trunk (capillary malformation)

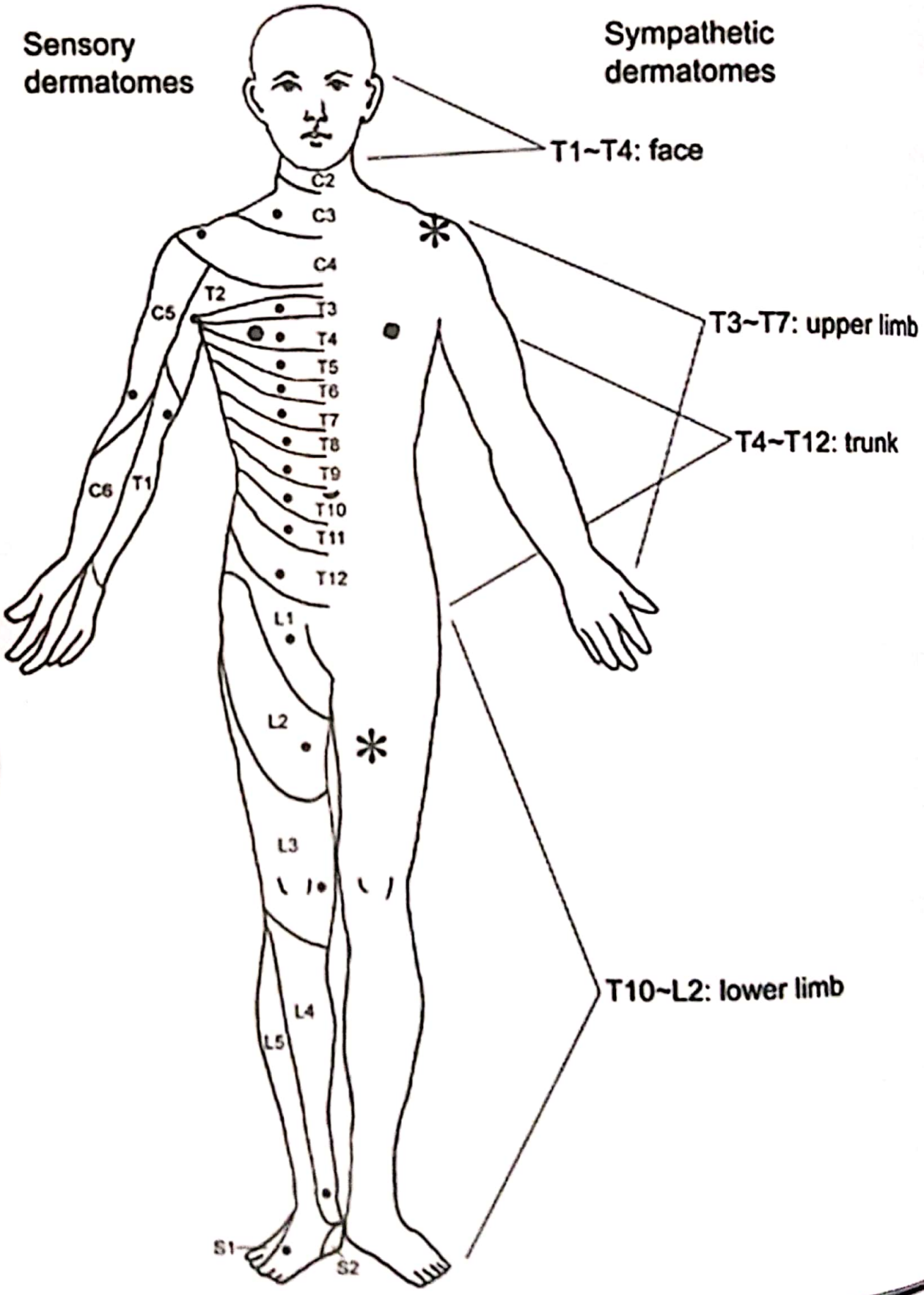
STEP V: MOTOR SYSTEM

See Motor system exam for details

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry & progressive centile charts



DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old **emaciated sick looking** child who is **conscious** and **cooperative** with IV cannula in place.
Gait could not be assessed as the child is unable to walk.
Bulk is bilaterally reduced in all the limbs; **tone is increased; reflexes are exaggerated** & **plantars are up going. Superficial reflexes are present/Absent at level** ___.
Sensory system is intact/There is sharp sensory level at _____ (Dermatome Range)
Spine is normal, **bladder** is not palpable and there are no **bed sores**.
BCG scar is absent

DIFFERENTIALS

1) **Pott's disease/TB spine**
2) **SOL spine** (tumor, abscess) .... There will be complete paralysis with down going plantars
3) **Transverse myelitis** (Sharp sensory level is present)

	INVESTIGATIONS	TREATMENT
Pott's disease	CBC, ESR Mantoux/ Quantiferon gold test X ray spine, Chest X ray MRI spine	ATT for 1 yr Jacket for stabilization of spine
Spinal tumor	MRI spine	Surgical resection Grade III & IV Astrocytoma can't be resected so radiotherapy and chemotherapy are given

TIPS & TRICKS

	Site of Lesion	Effect
C8	Above C8	Spastic Quadriplegia
C8-T1	C8-T1	UL: LMN signs LL:UMN signs
T	Thoracic region	Spastic paraplegia (With spared UL)
T10	Below T10	Absent superficial abdominal reflexes
LS	Lumbosacral	Flaccid paraplegia (With spared UL)

NOTES

SPASTIC PARAPLEGIA

1) General Look
2) Gait + Back
3) Motor system
4) Sensory exam
5) Relevant GPE

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient [Lying on couch/parent's lap]**
d. **Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)**
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Movement of limbs, Sick or well looking

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'* (Most likely child will be unable to walk)

1) *Tilting to a side*: Examine spine for MMC,scar, tenderness, mass etc.
2) Look for patulous anus
3) Look for bed sores

STEP III: MOTOR SYSTEM

See Motor system exam for details (Don't miss bladder & superficial reflexes)

STEP IV:SENSORY SYSTEM EXAM

Along dermatomes to see sensory level

RELEVANT GPE

Hands	Pallor
Pulse	As part of vitals
Temperature	To rule out febrile illness
BCG scar	To rule out TB spine
Chest	For crepts/aspiration

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) Fundoscopy (extradural tumors can block CSF)

Complications: Hydrocephalus, epilepsy(GTC & complex partial fits), brain tumour, increased risk of ALL,WT/Pheochromocytoma, Rhabdomyosarcoma, Precocious/delayed puberty

TIPS & TRICKS

1) If command is GPE then do Relevant GPE first and Motor system after that; But if command is Motor system then do Motor system first then relevant GPE.
2) **NF/Von Recklinghausen disease**: It occurs due to an abnormality of neural crest differentiation & migration during the early embryogenesis. **(Autosomal Dominant)**

NF1: 2 out of 7 criteria required:

1	**Café au lait spots**	At least 6 in number (> 5mm in pre-pubertal & > 15 mm post pubertal) May be present at birth? obvious by 1 year Present in 100% cases (spare the face ; found on trunk and extremities)
2	**Axillary & inguinal freckling**	> 2-3 mm hyperpigmenetd area Freckling appears in mid childhood
3	**Plexiform neuroma**	May involve blood vessels, GIT, skin, or peripheral nerves Skin over plexiform neuroma is more pigmented than café au lait spots. They are rubbery lesions with purpulish discoloration of skin. They can cause overgrowth of extremity and bone deformity. Due to hormonal influence they appear at puberty.
4	**Lisch nodules**	2 **or more lisch nodules** on slit lamp examination (rarely seen by naked eye) >75%in NF1 (100 % by 21 yr), Absent in NF2
5	**Optic glioma**	Visual acuity disturbance Afferent pupillary defect (on swinging light test) : Light in affected eye causes pupillary dilatation rather than constriction
6	**Sphenoid dysplasia:** Pulsating proptosis	
7	**1st degree relative**	

NF2:10%

1) Confirmed (definite) diagnosis of NF2:

Bilateral vestibular schwannomas (acoustic neuroma)

2) (Probable) diagnosis of NF2:

Family history of NF2 **AND** unilateral vestibular schwannomas or any 2 of the following tumor types: meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity, juvenile cortical cataract

NOTES

Ears	Decreased hearing (NF2)

STEP V: ABDOMEN

1) **Abdominal mass** (large visceral fibroma at puberty)
2) **Tanner staging** (Delayed or Precociuos) *(Seek permission before checking)*

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry (if incomplete)
3) Fundoscopy (Increased ICP)
4) Formal visual & audiological assessment
5) Slit lamp exam for Lisch nodules

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
His **gait** and **spine** are normal.
FOC is ____cm (appears large for age but I would like to plot it on centile charts). **Height** is __ cm (appears below 3rd centile with a short upper segment) and **weight** is ___kg.
HR is __/min, **RR** is __/min & **B.P** is ___mmHg (appears above 95th centile but I would like to plot it on centile charts)
There was no **neurological deficit**.
There are **multiple hyper pigmented spots** over the trunk maximum measuring 10mm in size .
There is **axillary & inguinal freckling**.
Eye examination was unremarkable and **hearing** was intact.
No **mass** was palpable in the abdomen and **Tanner staging** was pre-pubertal.

DIFFERENTIALS

1) NF-1
2) NF-2
3) Legius syndrome
4) LEOPARD syndrome

INVESTIGATIONS

1) **NF1 gene mutation**
2) **MRI Brain** (focal areas of demylenation)
3) **Slit-lamp examination** for Lisch nodules

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. PARENTAL COUNSELLING

II. SUPPORTIVE TREATMENT

Treatment of fits, hydrocephalus, genetic counselling

NEUROFIBROMATOSIS (NF)

1) General Look
2) Gait + Back + Anthropometry
3) Motor System
4) Relevant GPE
5) Abdomen

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For slurring/poor articulation/intellectual impairment)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **P**osition patient [Lying/Sitting on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Facial asymmetry due to facial neurofibroma
Respiratory rate

STEP II: GAIT , BACK & ANTHROPOMETRY

Ask: *'Bachaa Chal saktaa hai?'*

1) **Gait:** Limp due to CVA secondary to HTN
2) **Spine**: scoliosis, spinal neurofibroma
3) **FOC** : Macrocephaly (Brain tumor/Hydrocephalus/Aqueductal stenosis)
4) **Height**: Disproportionate short stature (US:LS = Low; due to scoliosis)

STEP III: MOTOR SYSTEM

See Motor system exam for details

STEP IV: RELEVANT GPE

Skin	Screen skin for café au lait spots > 6 of > 5mm are significant Measure maximum size & number of café au lait spots (Appear at birth- 1 year) Scratch marks (due to pruritis) Axillary & inguinal freckling(appears in mid childhood)
Pulse	As part of vitals
BP	HTN (essential , CoA, Renal artery stenosis, Pheochromocytoma)
Eyes	Proptosis, Decreased visual acuity (optic glioma) Ptosis (Plexiform neuroma of eye lid) Strabismus, Extra ocular movements, Lisch nodules

Treatment of Cardiac, endocrine, GI, Ocular complications
Physiotherapy and Orthopedic treatment of contractures

TIPS & TRICKS

1) Distal distribution of muscle wasting in myotonic dystrophy is an exception to the general rule of myopathies.

NOTES

Abdomen: Masses, constipation

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry
3) Functional assessment

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
He has **narrow head; inverted V shaped upper lip** and **scalloped concave temporalis muscle.**
Speech, gait and **spine** are normal. **Gower's sign** and **winging of scapula** are present.
Muscle bulk, Tone and **power** are reduced distally.
Deep tendon reflexes are diminshed.
There is **thenar, hypothenar, interossei muscle wasting** .
FOC is _____cm (I would like to plot it on centile charts)
BCG scar is present.
He has **high arched palate** but no evidence of **cataract, External opthalmoplegia, Thyromegaly** or **Sternocleidomastoid (SCM) muscle wasting.**

DIFFERENTIALS

1) Myotonic muscular dystrophy
2) Congenital myopathies
3) Atonic CP
4) Hypothyroidism
5) GBS

INVESTIGATIONS

I. FOR DIAGNOSIS & SUPPORTIVE

1) **CPK** (? in hundreds)
2) **EMG** (Helpful in toddlers/early school age)
3) **Mutation analysis** (CTG repeat at Chromosome 19)
4) **Muscle biopsy** (degeneration with little fibrosis)

II. TO RULE OUT COMPLICATIONS

1) **ECG** (For Arrythmias, Heart block)
2) **Chest X-ray** (For Aspiration)
3) **Endocrine assessment** (TFTs, Cortisol/Aldosterone levels for adrenal function, OGTT)
4) **Immunoglobulin levels** (?IgG)

TREATMENT

I. MDT (Multi-disciplinary team approach)

II. SUPPORTIVE TREATMENT

Phenytoin/Carbamazepine/Mexiletine/Procainamide/Quinidine sulphate (? Myotonia)

MYOTONIC CHILD

1) General Look
2) Gait + Back
3) Motor system
4) Relevant GPE
5) CVS, Abdomen

WIPER

a. **W**ash your hands with sterilizing solution/Warm your hands
b. **I**ntroduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For slurring/poor articulation/intellectual impairment)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **P**osition patient [Lying/Sitting on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Dysmorphism, narrow head, inverted V shaped upper lip, Scalloped concave temporalis muscle

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*
1) Gower's sign
2) Winging of scapula
3) Back

STEP III: MOTOR SYSTEM

See Motor system exam for details

STEP IV: RELEVANT GPE

Hands	Thenar, hypothenar, interossei muscle wasting
Pulse	As part of vitals
BCG scar	For Immunization status
FOC	To rule out CP
Eye	Cataract, External opthalmoplegia
Oral cavity	Thin & atrophic Tongue, High arched palate
Thyroid	Goitre
SCM	For wasting

STEP V: CVS & ABDOMEN

CVS: Block, Arrythmias

VII. Rituximab, a monoclonal antibody to the B-cell CD20 antigen.
For refractory patients

NEONATES WITH TRANSIENT MATERNALLY TRANSMITTED MYASTHENIA GRAVIS

Cholinesterase inhibitors for only a few days or occasionally for a few weeks, especially to allow feeding. (No other treatment is usually necessary)

NOTES

Mostly have permanent disease extending into adult life

Cure

Immunosuppression, thymectomy, and treatment of associated hypothyroidism

Genetically determined congenital myasthenic syndromes may show initial worsening in infancy but then remain static throughout childhood and into adult life.

Advice

Avoid Aminoglycoside antibiotics

Consult before surgery (**neuromuscular blocking drugs**)

2.SPECIFIC THERAPY

Mild myasthenia gravis : No treatment required

I. Cholinesterase-inhibiting drugs (Primary therapeutic agents)

IM	**Neostigmine Methylsulfate** (0.04 mg/kg) may be given IM every 4-6 hr
ORAL	Oral **Neostigmine bromide**, 0.4 mg/kg every 4-6 hr (30 min before meals to improve swallowing in case of dysphagia as major problem)
	Pyridostigmine is an alternative; the *dose required is approximately 4 times greater than that of neostigmine*, but it may be slightly **longer acting**.
Overdose	Cholinergic crises; Atropine blocks the muscarinic effects but does not block the nicotinic effects that produce additional skeletal muscle weakness.
No role	Familial myasthenia gravis caused by **absence of end plate AChE**, cholinesterase inhibitors are not helpful and often cause increased weakness; these patients can be **treated with ephedrine or diaminopyridine**, both of which increase ACh release from terminal axons.

II. Long-term steroid treatment with prednisone

Because of the autoimmune basis of the disease

III. Thymectomy

Most effective in:

1) Patients who have high titers of anti-ACh receptor antibodies
2) Symptomatic for <2 yr

Thymectomy is **ineffective** in *congenital and familial forms* of myasthenia gravis.

IV. Treatment of hypothyroidism

Usually abolishes an associated myasthenia without the use of cholinesterase inhibitors or steroids.

V. IV immunoglobulin

Beneficial and should be tried before plasmapheresis because it is less invasive.

VI. Plasmapheresis

Effective in those who do not respond to steroids

BUT plasma exchange therapy provides only temporary remission

Plasmapheresis and IV immunoglobulin appear to be most effective in patients with high circulating levels of anti-ACh receptor antibodies.

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious & cooperative** with an IV cannula in place.
He has bilateral **ptosis** but no dysmorphism.
He is **unable to sustain upward gaze**; **head elevation above table** and **arm elevation above head**. He developed **rapid fatigability of muscles** on repetitive opening n closing of hands.
Tone, power, reflexes are normal in both upper & lower limbs.
All the accessible **cranial nerves** are intact & there is no **myotonia** or evidence of **aspiration**.

DIFFERENTIALS

1) Autoimmune myasthenia gravis
2) Congenital myasthenic syndromes
3) Toxin-induced myasthenia (Botulism)
4) Organophosphate poisoning

INVESTIGATIONS

I.FOR DIAGNOSIS

1) **EMG** *(Decremental response to repetitive nerve stimulation)*
2) **Serological studies**
Plasma Anti-AChR antibodies (inconsistently demonstrated, 30% of affected adolescents)
Anti–muscle-specific tyrosine kinase (MuSK) **circulating antibodies** (Seen in kids with negative titers of AChE exhibit)
Anti-rapsyn, anti-Dok7, COLQ, and ChAT antibodies (congenital myasthenia gravis)
3) **Tensilon test**: **Administration of a short acting cholinesterase inhibitor** (edrophonium chloride= Tensilon) . Ptosis &ophthalmoplegia improve within few seconds, & fatigability of other muscles decreases.

<2yr old	>2 yr old
Prostigmin methylsulfate (Neostigmine)	Edrophonium chloride (Tensilon)

4) **Muscle biopsy** (limited role)

II.SUPPORTIVE

1) **Antinuclear antibodies** (to rule out other autoimmune diseases)
2) **Test for abnormal immune complexes** (to rule out other autoimmune diseases)
3) **Serum creatine kinase level** (CPK) normal in myasthenia gravis
4) **Electrocardiographic findings** (Normal as heart is not involved)

III.TO RULE OUT COMPLICATIONS

1) **Radiographs of the chest**
May reveal an enlarged thymus, but the hypertrophy is not a thymoma
2) **Tomography / CT or MRI of the anterior mediastinum** : To delineate thymoma

TREATMENT

MDT: Pediatrician, Rehab medicine specialist, Pediatric Neurophysician

1. PARENTAL COUNSELING /PROGNOSIS

Spontaneous remission after a period of months or years (Some patients with autoimmune MG)

MYASTHENIA GRAVIS

1) General Look
2) Maneuvers for Myasthenia Gravis
3) Motor system examination
4) 180 degree manoeuvre
5) Chest

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building (*Interact with child, assess speech for dysarthria*)
c. **P**osition patient [Make him lie on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Bilateral ptosis, myopathic facies

STEP II: MANOEUVRES FOR MYASTHENIA GRAVIS

1) Ptosis worsens when the patient is asked to sustain an upward gaze for 30-90 sec.
2) Patient cannot elevate his head above examination table for > few sec.
3) Patient cannot elevate his arms above head for more than 1-2minutes.
4) Repetitive closing & opening of hands produces fatigability of muscles.

STEP III: MOTOR SYSTEM EXAMINATION

See Motor system exam for details

1) Tendon reflexes may be diminished but rarely lost.
2) Involvement of limb girdle & distal muscles
3) Check for myotonia
4) Check extraocular movements

STEP IV: 180 DEGREE MANOEUVRE

For young child or infant: For hypotonia, poor head control

STEP V: CHEST

Auscultation for aspiration

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Anthropometry

b) Acquired hydrocephalus (e.g. meningitis): Normal growth before insult
c) Congenital problem (e.g. aqueduct stenosis): Earlier deviation from the norm

1) Hemiplegia with hydrocephalus: Postmeningitic sequel. Subdural hematoma
2) MMC short case is similar to Large head short case except examine & describe swelling of MMC.

NOTES

INVESTIGATIONS

1) **CBC** to rule out anemia and infection
2) **Plain x ray Skull** (increased convolutions/silver beaten appearance; increased separation of sutures, erosions of posterior clinoid)
3) **USG cranium**
4) **CT scan/MRI brain**
5) **TORCH screening**
6) **CSF/Ventricular tap to rule out infection**
7) **Auditory assessment**
8) **Visual assessment**

TREATMENT

HYDROCEPHALUS

1) **MDT**
2) **Counselling of parents**
3) **Treatment of acute problems ± Antibiotics**
4) **Medical treatment** (Acetazolamide ± diuretic)
5) **Surgical treatment** (VP shunting, ETV/endoscopic third ventriculostomy which doesn't require repeated shunting)

MENINGOMYELOCELE

1) **Surgery**: MMC repair is urgent if CSF leak is there otherwise a few days after birth
2) **CSF diversion**: VP shunting
3) **Associated anomalies**: Club foot, renal anamolies
4) **Bladder training**: Intermittent catheterization, latex free catheter and gloves, periodic RFTs. Urine R/E & CS, MCUG, or artificial sphincter implantation.
5) **Bowel training** : Timed enemas for rectal dis-impaction
6) **Ambulation aids, braces**
7) ***Prevention:*** *0.4 mg folic acid to all women 1 month before pregnancy, 4 mg in high risk mother*

TIPS & TRICKS

1) Interact while palpating watching eye movements & responsiveness.
2) **Fontanelle closure**

Posterior : 4 months	Anterior : 18 months

3) Check Lower limbs before the upper limbs (as LL are first affected in hydrocephalus, because the tracts supplying them run closer to the ventricles)
4) Three Must do areas : Head , Back, Eye movements [particularly the upward gaze (for Parinaud syndrome) and lateral rectus function (for raised intracranial pressure compressing the sixth nerve)]
5) On Growth charts
   a) Measurements crossing percentile lines = significant pathology (Except 'familial large head' in which initial deviation & then stabilization occurs after 2 yrs of age)

There is **no respiratory distress, dysmorphism, skeletal anomalies** or **neurocutaneous stigmata**.
He is/not **moving his lower limbs**.
Head is **enlarged symmetrically** with **shiny skin and prominent veins**.
Spine and anus are normal.
FOC is 50 cm which is > 2SD above mean but I would like to plot it on a centile chart.
Anterior fontanelle is wide open, non-pulsatile, normotensive, measuring 7 ×3 cms. **Posterior fontanelle** is open measuring 1x1 cm. Sutures are wide open.
Cracked pot sign is present but there is no **bruit**.
He has **sunset sign** but no evidence of **cataract, squint or nystagmus**.
Hearing appears normal.
A **VP Shunt** is placed on right side and is **patent** and traceable till abdomen.
Rachitic rosary is not present.
Both heart sounds are normal with no **murmur**.
Abdomen is soft with no visceromegaly.
Lower limb has LMN signs while **upper limb** examination is normal.
Gait: _____ **Cerebellar signs** are/not present.
No evidence of **pallor, jaundice, petechaie, bruise** or **clubfoot**.

DIFFERENTIALS

CAUSES OF HYDROCEPHALUS

Communicating	Non communicating/Obstructive
1. Pyogenic Meningitis 2. TBM 3. TORCH infections 4. Choroid plexus papilloma	1. Congenital Aqueductal stenosis 2. Dandy Walker 3. Arnold chiari malformation 4. Holoprosencephaly 5. Porencephalic cyst 6. Posterior fossa tumors

CAUSES OF LARGE HEAD

Large ventricles / subarachnoid spaces	Large brain (megalencephaly)
1. Obstructive/non-communicating hydrocephalus e.g. aqueduct stenosis or posterior fossa tumours 2. Failure of CSF absorption/communicating hydrocephalus e.g. meningeal adhesions after meningitis 3. Overproduction of CSF/communicating e.g. choroid plexus papilloma	1. **Sotos syndrome** (cerebral gigantism) 2. **Neurocutaneous syndromes:** (a) Neurofibromatosis type 1 (NF-1) (b) Tuberous sclerosis (TS) (c) Klippel–Trenauney–Weber syndrome (KTW) (d) Sturge–Weber syndrome (S–W) 3. **Inherited metabolic disorders:** (a) Lipidoses (e.g. Tay–Sachs disease) (b) Mucopolysaccharidoses (MPS). (c) Leukodystrophies;(Alexander disease)
Large bones	**Large brain (localised enlargement)**
1. Achondroplasia 2. Rickets 3. Osteogenesis imperfecta (OI) 4. Chronic haemolytic anaemias	1. Cerebral tumours (glioma or ependymoma) 2. Cerebral abscess
	Large bleed
	Subdural haematoma (unilateral or bilateral)

	2) **Anterior & Posterior Fontanelle** measurements 3) **Measure MMC** dimensions
Palpate	**For suture separation** **Fontanelle patency, pressure, pulsations** *(Split sutures & absent ant. fontanelle pulsation is a sign of hydrocephalus)*
Percuss	**Crack pot sign/Macewan sign** (Tapping with finger for resonant sound at junction of frontal,temporal and parietal bone)
Auscultate	**Auscultate** (Both temporal fossae, both eyeballs and both retroauricular regions) *For evidence of an AV malformation involving the great vein of Galen.*

STEP III: EYES & EARS

1) Visual acuity, External ocular movements, squint
2) Pupils, cataract (toxoplasmosis)
3) Hearing (paper/wrapper)

Severe frontal bossing can be a visual barrier, (false impression of an upward -gaze palsy)

STEP IV: VP SHUNT →CHEST →CVS→ABDOMEN

1) **Trace shunt** (bilaterally) & **check patency**
2) **Chest** : Scars (VA shunt), Signs of rickets
3) **CVS** : Auscultate heart for murmur (TORCH)
4) **Abdomen**: Scars (VP shunt), Hepatosplenomegaly (MPS)
5) **Bladder** (neurogenic bladder)

STEP V: MOTOR SYSTEM EXAMINATION (LL before UL)

See Motor system exam for details
UMNL signs : (Hydrocephalus, intracranial tumour)
LMNL signs : (Spina bifida, leukodystrophies)
Cerebellar signs (Dandy-Walker syndrome) + Gait (Perform in elder child)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Measure the parents' heads (if no obvious cause is known)
2) Request the progressive percentiles of the child
3) Blood pressure & heart rate
4) Fundoscopy
5) Testing sensation (e.g. sensory level in spina bifida)
6) Formal visual & audiological assessment
7) Developmental assessment

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child of thin lean built who is **conscious** but I **could not gain full cooperation.**
He has a **cannula** on dorsum of left hand & **interacting appropriately** for his age.

LARGE HEAD

1) General Look
2) Head & Back
3) Eyes & Ears
4) VP shunt →Chest →CVS→Abdomen
5) Motor system examination

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building (*Interact with child, assess speech*)
a. **P**osition patient [Make him sit on couch/parent's lap]
b. **E**xposure: (Ideal exposure in male child: Cap off, Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
c. **R**ight side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) **Alertness**
2) **Respiratory rate**
3) **Movements** (hypotonia, poor head control in Tay–Sachs, Hemiplegia in SDH, paraplegia in spina bifida)
4) **Dysmorphic features** (e.g. Sotos, MPS), Facies of hemolytic anemias
5) **Skeletal anomalies** (e.g. achondroplasia, OI)
6) **Skin** (Neurocutaneous stigmata) (Bruising in NAI)
7) **Growth parameters** (tall = Sotos ; short= achondroplasia) (FTT: e.g. Tay–Sachs, subdural from NAI, congenital toxoplasmosis)

STEP II: HEAD & BACK (Make child sit with support)

<table>
<tr><td>Inspect</td><td>1) Signs of hydrocephalus ('sun-setting' of the eyes, Prominent scalp veins, shiny skin)
2) Shape: Assessed from all angles, and is best described in terms of 'anteroposterior diameter' and 'biparietal diameter', and 'frontal bossing' and 'occipital prominence'.
<table>
<tr><td>Frontal prominence: obstructive hydrocephalus</td><td>Parietal prominence: subdural fluid/ porencephalic cyst</td></tr>
<tr><td>Occipital prominence: Dandy walker</td><td>Small post fossa: Aqueductal stenosis</td></tr>
</table>
3) Look for titubation/nodding of head or trunk, or truncal ataxia from Dandy–Walker syndrome
4) Look at back at this point (while inspecting occiput)
a) MMC, swelling, Tuft of hair, Midline scar (spina bifida)
b) Scoliosis (associated spina bifida,NF-1)
c) Patulous anus</td></tr>
<tr><td>Measure</td><td>1) FOC : 3 times (Maximum is considered)</td></tr>
</table>

Serum & urine amino acids/organic acids (For IEM) 6. **Urine mucopolysaccharide, skeletal survey, Enzyme assays** (For Storage disorders) 7. **Very long chain fatty acids** (? in Zellweger syndrome)	4. **SMN 1 gene** (for SMA) 5. **Muscle biopsy**

TREATMENT

MDT

Supportive care

Physiotherapy, nutritional rehabilitation, occupational therapy

Identification & treatment of treatable disorders

Thyroxine	Hypothyroidism
Pyridostigmine	Congenital myasthenic syndromes
Specific dietary modifications	Metabolic disorders
Gene therapy	SMA

Evaluate for associated cardiac dysfunction

Genetic counseling (in case of hereditary disorder)

TIPS & TRICKS

1) Be opportunistic in examining infants.
2) Signs of UMNL make it floppy strong and signs of LMNL make it floppy weak.

NOTES

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Anthropometry (if Not done)
2) Fundoscopy (if relevant)
3) Primitive reflexes (if CP)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ months old infant who is **conscious** but I **could not gain full cooperation**.
He is having **thin built** (his weight seems less for his age) and a **cannula** in his ___ forearm.
He is **floppy weak** (peripheral hypotonia) with **alert/dysmorphic facies**, **frog leg/hypotonic posture** with/without **tongue fasciculation.**
He has **weak cry** with/without **respiratory distress** in the form of tachypnea, subcostal recessions and nasal flaring.
He is **afebrile**; **pulse** is 100 bpm, regular in rhythm and normal in volume and character; **Respiratory rate** is 50/min.
His **length** is__cm; **weight** __kg and **FOC** __cm but I would like to plot these on centile charts.
Muscle bulk is reduced in all limbs and he has **signs of lower motor neuron lesion** in the form of hypotonia, power 1/5, absent deep tendon reflexes and plantars_____
He **cannot hold his neck**, sit without support and sags during ventral suspension.
Spine is normal.
Abdomen is protuberant with no visceromegaly.
Both **heart sounds** are audible with no murmur.
No evidence of **myopathic facies**, **ptosis** (Mysthenia Gravis), **Cataract**, **drooling** (Botulism), **petechaie** (TORCH), **aspiration**, **contractures or primitive reflexes** (C.P), **umbilical hernia** (Hypothyroidism), **Hepatomegaly** (GSD) or **Undescended testes**

DIFFERENTIALS

FLOPPY STRONG/CENTRAL HYPOTONIA	FLOPPY WEAK/ PERIPHERAL HYPOTONIA
1. Cerebral palsy 2. Cerebral malformations 3. Hypothyroidism 4. TORCH infections 5. Down/Prader willi Syndrome (if dysmorphic) 6. IEM 7. Storage disorders (Taysach, MPS, ALD)	1. SMA type 1 2. Congenital myopathies 3. NMJ disorders (Myasthenia gravis, Botulism)

INVESTIGATIONS

FLOPPY STRONG	FLOPPY WEAK
1. **MRI brain** (for CP/Cerebral malformations) 2. **Thyroid function tests** (for Hypothyroidism) 3. **TORCH screening** (for TORCH infections) 4. **Chromosome analysis** (for trisomy 21, Prader-Willi syndrome) 5. **ABGs , Serum and CSF lactate, Serum ammonia,**	1. **Creatinine kinase** (CPK) 2. **Lactate dehydrogenase** (LDH) 3. **Electromyography, repetitive nerve stimulation test**

FLOPPY INFANT

1) General Look
2) Motor system examination
3) 180° maneuver
4) Relevant GPE

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Make him sit on couch/parent's lap]
d. Exposure: (Ideal exposure in infant: Shirt & trousers off: Examine in nappies) [Seek parent's help to undress] (Beware of hypothermia, Be opportunistic)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Look for dysmorphism (Down's Syndrome), Myopathic facies (inverted V shape, shallow temporal fossa, ptosis)
2) Posture
3) Respiratory rate & pattern

STEP II: MOTOR SYSTEM EXAMINATION

See Motor system exam for details

STEP III: 180 DEGREE MANEUVER

See Developmental assessment for details
(Examine back during this maneuver)

STEP IV: RELEVANT GPE

1	Pulse rate	For Hypothyroidism
2	FOC (3 times)	For CP
3	Anterior fontanels	Wide open (Down's, Hypothyroidism)
4	Eyes	Cataract (TORCH)
5	Mouth	Tongue fasciculation (SMA)
6	CVS	Cardiomyopathy (GSD), PDA (TORCH)
7	Chest	For Aspiration
8	Abdomen	For umbilical hernia (Hypothyroidism), Hepatomegaly (GSD), Undescended testes
9	Back	Spine, Anal tone (If Not examined during 180° maneuver)
10	Anthropometry	Length, Weight

Redress the child and say thank you!

Neuroimaging

- If focal sign
- No recovery in 1 month
- If tumor is suspected

TREATMENT

MDT (Multi-disciplinary team approach)
Counsel the parents
Treatment

- ✓ Oral prednisone (1 mg/kg/day for 5-7 days, followed by a 1 wk taper) started within the first 3 days after onset.
- ✓ Oral Acyclovir (±)
- ✓ Protection of the cornea (methylcellulose eye drops/ocular lubricant especially at night)
- ✓ Physiotherapy
- ✓ Surgical decompression

PROGNOSIS

Complete spontaneous recovery within a few weeks from onset. (90%)
Residual facial weakness (<10%)
Recurrent episodes of facial weakness (15%)
No recovery within few weeks: Do NCS of facial nerve
(Suspect facial nerve schwannoma, infiltration by leukemin cells.

TIPS & TRICKS

1) A facial nerve palsy may be congenital; idiopathic **(Bell palsy);** or secondary to trauma, demyelination (Guillain-Barré syndrome), infection (Lyme disease, herpes simplex virus, HIV), granulomatous disease, neoplasm, or meningeal inflammation or infiltration.
2) Facial nerve lesions that are proximal to the junction with the chorda tympani will result in an inability to taste substances with the anterior two thirds of the tongue.
3) If necessary, taste can be tested by placing a solution of saline or glucose on one side of the extended tongue. Normal children can identify the test substance in <10 sec.

NOTES

Cranial Nerve 8

1) Whisper test
2) Rinnie test
3) Weber test

STEP IV: REST OF THE CRANIAL NERVES (To know extension)

Visual fields	2nd
External eye movements (Make H)	3rd ,4th ,6th
'Clench your teeth tight': feel the muscle bulk on each side	5th
Asymmetry of palate elevation	9th
Asymmetry of shoulder shrugging	11th
Tongue deviation	12th CN

STEP V: MOTOR SYSTEM EXAMINATION

See Motor system exam for details (include Gait)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Otoscopy (for vesicles in Ramsay Hunt syndrome)
2) Examination for taste sensations (If Not done)
3) Blood pressure (For Brain stem Gliomas)
4) Fundoscopy (if relevant)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious & cooperative** with normal speech.
He has Left sided **Lower Motor Facial Nerve palsy** as evident by inability to frown and close the eye. Disappearance of nasolabial fold, drawing of the mouth to the unaffected side.
Hearing is intact/decreased on __side.
There is no **ptosis**, or involvement of other **accessible cranial nerves.**
Motor system examination including **Gait** is normal. There is no **myotonia.**
Bp is 100/60 mmHg.

DIFFERENTIALS

LMN type Facial palsy		UMN type Facial palsy	
Unilateral	Bilateral	Unilateral	Bilateral
Bell's palsy Trauma Infection (Ramsay Hunt syndrome) Congenital lesion	Myasthenia gravis GBS (Miller Fisher) Myotonic dystrophy	Stroke Trauma	Cerebral palsy

INVESTIGATIONS

CBC (To Rule out evidence of infection)

FACIAL WEAKNESS

1) Inspection
2) Talk to child
3) Examination of CN 7, 8
4) Other CN
5) Motor system examination

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
c. Position patient [Make him sit on couch/parent's lap]
d. Exposure: (as required) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: INSPECTION

Look for asymmetry of face, any involvement of limb, neurocutaneous stigmata

STEP II: TALK TO CHILD

1) *Beta apka naam kia hai?* (For hoarseness/slurring/Hearing assessment)
2) *Kis class main parhtay hain?* (For Mental age)

STEP III: EXAMINATION OF CN 7 & 8

Cranial Nerve 7

'Look up and raise your eyebrows'	It tests the frontalis and is useful in differentiating UMNL (upper facial muscles are preserved due to bilateral innervation) from LMNL, where the upper facial muscles are affected
'Close your eyes tightly'	Compare both sides, noting any asymmetry between the degree the eyelashes are buried on either side. This may detect an obvious **Bell's phenomenon**, where there is rolling upward of the eyeball when attempting to shut the eyes forcefully, as eye closure may not be possible in lower motor neuron lesions.
'Keep them shut; stop me opening them'	This tests the orbicularis oculi muscles
'Now show me your teeth'	Allows assessment of any asymmetry of the nasolabial grooves. The **mouth will be drawn towards the normal side** if there is a unilateral lesion of either upper or lower motor neuron type
'Puff out your cheeks; keep them like that'	Demonstrate this and then tap with your finger over each cheek to detect ease of air expulsion on the affected side.
Check Taste sensations	Anterior 2/3rd of tongue

TIPS & TRICKS

1) If command is motor system examination then do motor system first and then GPE & relevant systemic exam.
2) Dystonia = Sustained muscle contraction

Increase in Dopamine causes Chorea	Decrease in Dopamine causes Dystonia
(Increased Dopa causes dancing)	*(Decreased Dopa causes brakes in body)*

NOTES

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious but poorly interactive** with Dystonia involving Right arm.
There is no **dysmorphism, deformity, neurocutaneous stigmata.**
Gait is _____ and **spine** is normal.
Muscle bulk is normal and symmetrical in all limbs with no scar marks or obvious deformity.
Tone is decreased/normal, **power** grade 5/5 with normal **DTR** and **plantars.**
All accessible **Cranial nerves** are intact and **coordination** is present/absent.
His FOC is _____cm (Which appears appropriate for age but I would like to plot it on centile charts)
He is **afebrile** and there is no evidence of **Clubbing, palmar erythema, Pallor, Jaundice, Spider nevi, Edema, Hepatosplenomegaly, Ascites or heart failure.**

DIFFERENTIALS

1) Wilson's disease
2) Drugs (Dopamine antagonists: Haloperidol, Metocloperamide, Risperidone, Antipsychotics)
3) Cerebral palsy (Post Kernicteric/Post Asphyxial)
4) SSPE
5) Post Hemiplegic
6) Primary inherited dystonia (e.g. Segawa disease)

INVESTIGATIONS

I will investigate as per possible differentials

For Wilson's disease	LFTs, Serum ceruloplasmin level, 24 hr urinary copper level, Slit lamp examination for KF rings, USG Abdomen, MRI Brain
For Drug induced	Drug levels
For CP	MRI Brain
For SSPE	Anti-measles antibodies

TREATMENT

After establishing my diagnosis by detailed history and clinical exam:

1) **Multidisciplinary team approach**
2) **Counseling of Parents**
3) **Treatment of acute problem** *(Tip: Parkinsonism like treatment)*

Anti cholinergics (For dysphagia)	Trihexyphenidyln2mg/day (Max 60-80mg)
Other drugs	Levodopa, Carbamazepine, Diazepam, Bromocriptine
Segmental dystonia	Botulism A injection
Generalized dystonia	Intrathecal Baclofen

4) **Treat underlying cause**

For Wilson's disease	Dietary restriction of nuts, chocolates, Pencillamine with zinc acetate
For CP	Anti-epileptics, Supportive care
Drug induced	Stop drug and start diphenhydramine
Surgery	Thalamotomy, Pallidectomy

DYSTONIA

1) General Look
2) Motor system
3) FOC
4) GPE
5) Abdomen, CVS, Chest

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For hoarseness/slurring)
   2) *Kis class main parhtay hain?* (For Mental age)
c. Position patient [Make him sit on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Look for dysmorphism, rash, Jaundice, Abnormal movements and its distribution

STEP II: MOTOR SYSTEM EXAMINATION

See Motor system exam for details (include Gait, Cranial Nerves, coordination)

STEP III: FOC

To rule out CP

STEP IV: GPE

Signs of CLD: Clubbing, palmar erythema, Pallor, Jaundice, Spider nevi, Edema (CLD)

STEP V: ABDOMEN, CVS, CHEST

1) Abdomen: Liver, spleen, ascites (CLD)
2) CVS, Chest (for signs of failure)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Fundoscopy
2) Slit lamp exam for KF rings
3) Anthropometry
4) Vitals

Walking or Stepping reflex	Birth ? 6 month	It is present at birth, though infants cannot support their own weight. When the soles of their feet touch a flat surface they will attempt to walk by placing one foot in front of the other. **This reflex integrates around 5–6 months as infants start attempting to walk after this reflex disappears.**
Galant reflex	Birth ? 6 month	When the skin along the side of an infant's back is stroked, the infant will swing towards the side that was stroked. **If the reflex persists past six months of age, it is a sign of pathology.**
Parachute reflex	6 months ? Lifelong	You can see this response when you hold the baby in a straight position under your armpits and turn him over on his belly quickly. He will extend his arms to break the fall.

NOTES

Primitive reflexes are present/absent.
His approximate overall developmental age is _____months. (Give range if Not sure)

TIPS & TRICKS

1) Usually an infant can be the case so be opportunistic.
2) Don't give eatables/bunties/saunf: He/She can choke upon it.
3) If child is able to perform a task, go to next relevant milestone in the same domain till you find a point where child can't perform e.g. if he can grasp a brick? give pellet to check pincer grasp
4) Ideally mention developmental age in each domain and then give approximate overall developmental age (usually derived from Gross/fine Motor age)
5) Perform just 2-3 primitive reflexes to touch upon it if time is short.
6) **Primitive reflexes**: *Set of transient reflexes evident in neonatal & post neonatal period*
   a) Controlled by most primitive parts of brain (i.e. Medulla & mid brain)
   b) Gradually suppressed by the development of the frontal lobes during first 3-12 months.

*(Described below in sequence of integration while doing **180 degree maneuver**)*

REFLEX	TIMING	METHOD & CLINICAL SIGNIFICANCE
Rooting reflex	Birth ? 4 month	A newborn infant will turn its head toward anything that strokes its cheek or mouth, searching for the object by moving its head in steadily decreasing arcs until the object is found. **It assists in the act of breastfeeding**
Asymmetrical tonic neck reflex (ATNR)	1 month ? 4 month	When the child's head is turned to the side, the arm on that side will straighten (extends) and the opposite arm will bend (flex). **Inability to move out of this position or the reflex continues to be triggered past 6 months of age : UMNL (e.g. CP)**
Symmetrical tonic neck reflex (STNR)	4-6 month ? 7-8 month	Place infant supine on couch. Passively flex and the extend head. On Flexion: Flexion of arms & extension of legs On Extension: Extension of arms & flexion of legs *(Aj's handy tip: arms go with position of head; legs do opposite)* **Persistence will prevent a child from crawling.**
Moro's reflex	Birth ? 4 month	Hold baby off the couch in supine position with your arms underneath their back. Lower the head and upper back suddenly (drop baby few inches). The legs and head extend while the arms jerk up and out with the palms up and thumbs flexed. **Asymmetric Moro's reflex: Fractured clavicle/Brachial plexus injury** **Bilateral absence of the reflex : Damage to the infant's CNS**
Palmar grasp reflex	Birth ? 5 month	When an object is placed in the infant's hand and strokes their palm, the fingers will close and they will grasp it with a palmar grasp. To best observe this reflex, on a bed where the child could safely fall onto a pillow, offer the infant two opposing little fingers (as index fingers are typically too large for the infant to grasp), and gradually lift. The grasp of it may be able to support the child's weight, they may also release their grip suddenly and without warning. The reverse motion can be induced by stroking the back or side of the hand.

Palmar grasp + mouths object: **6 month**	Can transfer object: 7 **months**
Picks pellet (immature pincer): **9 month**	Mature pincer grasp: **10-12 months**

1) **Gross motor: 180 degree maneuver**
   a. Lying supine (assess position adopted, e.g. Pithed frog: Hypotonic baby)
   b. Pull to sit by hands (to assess head lag and grasp)

Head lag disappears: **4 month**	Anticipated head lift before pull: **5 month**

   c. Sitting (assess sitting ability)

Head control: **4 month**	Sitting with support: **7 month**
Sitting without support: **8-9 month**	Pivot/Rotates trunk: **11 month**

   d. Hold up vertically, under axillae (Supported standing)

Bears most of weight: **6 month**	Stands holding furniture: **8.5 month**
Cruises around furniture: **10-11 month**	

   e. Ventral suspension

Lifts head in horizontal plane: **1.5 month**	Lifts head above body plane: **3 month**

   f. Place prone

Head turned to side+Pelvis higher than shoulders: **1 month**	Holds chin & shoulders off cot: **3 month**
Holds Chest & upper abdomen off cot + Rolls over prone to supine: **6 month**	Crawls: **9 month**

2) **Relevant**: FOC, Motor system exam, CVS (TORCH), Hepatosplenomegaly (Storage)
3) **Primitive reflexes** (Perform if asymmetry of movements of limbs in noted) *(see tips & tricks)*

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Primitive reflexes (if Not done)
2) Fundoscopy (if relevant)
3) Formal audiological & visual assessment (if needed)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child who is **conscious & cooperative** with no **dysmorphism, deformity, or abnormal movements.**
He is microcephalic with **FOC** of 39 cm.
He can Coo/Babble/say dada mama **(Language)**
Social smile is present; there is no stranger anxiety **(Social interaction)**
He can fix & follow up dangling ball to 180° **(Vision)**
He can turn head to rattle below level of ear **(Hearing)**
He has immature pincer grasp. **(Fine motor)**
On **180 degree maneuver** there is no head lag; he can sit with support; can bear his weight on holding vertically; can lift head above body plane on ventral suspension and can hold Chest & upper abdomen off couch on placing prone. **(Gross motor)**
Motor system, CVS and abdominal examinations are unremarkable.

DEVELOPMENTAL ASSESSMENT

1) General Look
2) Language/Social interaction
3) Vision
4) Hearing
5) Fine motor
6) Gross motor
7) Relevant
8) Primitive reflexes

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building
c. **P**osition patient [Make him sit on couch/parent's lap]
d. **E**xposure: (Expose as required)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

BASIC EQUIPMENT NEEDED

Color ball, Rattle, Blocks

1) **General Look** (Syndromic, Myopathic)
2) **Language/Social interaction**: *Ask mother to talk and play with child*

Cooing (Low soft gentle cry): **3 months**	Babbling (Unintelligible sound): **6 months**
Understands own name: **8 months**	Dada,Mama: **9 months**
First meaningful word: **1yr**	

Involve yourself in playing with child

Social smile: **1.5 months**	Laugh out loud: **4 month**
Stranger anxiety: **7 months**	Waves Bye Bye/Peek-a-boo: **10 months**
Give block & ask to give back: **11months**	

3) **Vision**: *Dangle 5cm colored ball at 25cm moving horizontally*

Fixes & follows up to 45° :**Newborn**	Fixes & follows up to 90° : **1.5 months**
Fixes & follows up to 180° :**3 months**	Can fix at 2cm block/ball : **4 month**
Can fix onto smarties: **8 months**	Can see distant object: **9 months**
If Child doesn't follow	
Shine light/Check papillary reflexes	

4) **Hearing**: While the child is busy looking at ball; Give rattle to attendant/examiner to make noise from behind at level of ear/below level of ear

Crying baby quietens: **1 month**	Turns head to sound at level of ear: **3 month**
Turns head below level of ear: **6 month**	Distraction test: **9 months**

5) **Fine motor**: *Give block/pellet to child* (check how he grasps)

Reaches for it: **4 month**	Reaches & gets it: **5 month**

II. SUPPORTIVE TREATMENT

Admit to ICU
Vitals monitoring
ABC
Treatment of acute conditions such as mastioditis/meningitis

Neuroprotection**

1) Control of temperature
2) Blood pressure
3) Glucose
4) Seizures

III. SPECIFIC THERAPY

Arterial Ischemic Stroke (AIS)	Cerebral sinovenous thrombosis	Hemorrhagic strokes (HS)
For secondary stroke prevention 1) Anticoagulants (IV heparin, subcutaneous LMWH, oral warfarin) 2) Platelet anti-aggregants (aspirin)	**Progressive CSVT** Anticoagulation	**Involve Pediatric Neurosurgeon** Surgery for ICB Surgery for AVM

V. FOLLOW UP/MONITORING OF DISEASE ACTIVITY

Long-term rehabilitation programs

NOTES

II. TO ASCERTAIN CAUSE

1) Blood: haematology
- FBC, ESR: polycythaemia
- Thrombophilia screen, fibrinogen: thrombophilia

2) Blood: biochemistry
- Electrolytes, magnesium
- Liver function tests
- CRP: inflammation
- Plasma lactate and CSF lactate: mitochondrial disorders
- Fasting glucose: diabetes
- Fasting lipid screen: hyperlipidaemias
- Thyroid function tests: Hashimoto thyroiditis/encephalopathy
- Ammonia: urea cycle disorders
- Homocysteine (free and total): methyltetrahydrofolate reductase (MTHFR) defi ciency can also be picked up by common mutation analysis on the thrombophilia screen, and if symptomatic has a raised plasma homocysteine
- Serum iron, total iron binding capacity, ferritin, red cell folate, and vitamin B12: iron deficiency and other nutritional disorders
- Plasma amino acids: aminoacidurias
- Carnitine (acyl, free, and total): B-oxidation defects

3) Urine: biochemistry
Urine organic and amino acids: homocystinuria, MTHFR defi ciency

4) Blood immunology and infection screen
- IgG, IgM, IgA: immunodeficiency
- Titres for infection screen of: Mycoplasma, Chlamydia, Helicobacter, Borrelia, Brucella; viruses (echo, Coxsackie, Epstein–Barr, Varicella, hepatitis B)
- ASOT, Anti DNAase B: streptococcal disease
- ANA, ANCA, anticardiolipin and antiphospholipid antibody: SLE and autoimmune disease

5) Imaging studies
- Echocardiogram & Doppler: endocarditis and other cardiac disease/ to detect a source of embolism
- Carotid Doppler studies to rule out dissection of the carotid arteries

MANAGEMENT & REHABILITATION

Goals of treatment

1) Limiting secondary neuronal injury	2) Prevention of future strokes

a) Tabulated Overview

MDT	Supportive R_x	Specific R_x	F/UP

b) Details

I. MDT (Multi-disciplinary team approach)

Pediatrician	P. Neurophysician	Physical therapist	Radiologist
Neurosurgeon	Ophthalmologist	Psychologist	Dietician

He has **Circumductive** (Hemiplegia) **gait** with negative/positive **Romberg sign**.
Spine is normal.
Muscle bulk is normal with no **fasciculations**.
Tone is increased in Rt Upper & lower limbs.
Ankle Clonus is illicitable in Rt foot.
Power is 2/5 in Rt Upper & lower limbs.
Deep tendon reflexes are exaggerated in Rt Upper & lower limbs.
Plantar reflex is upgoing in Rt foot.
Abdominal reflexes are intact and **bladder** is not palpable
There is **Right sided facial nerve palsy** while rest of the cranial nerves are intact.
FOC is _____cm (I would like to plot it on centile charts)
Signs of meningeal irritation are absent
There is/is no evidence of **involvement of Parietal lobe.**
There is/is no evidence of **pallor, clubbing, cyanosis, splinter hemorrhages, Radioradial/ Radiofemoral delay, lymphadenopathy, scars, shunts, cataract, murmur, hepato-splenomegaly,** or **edema.**
BCG scar is present.

DIFFERENTIALS

*Mnemonic: Stroke Says **STITCH ME***

Sytemic: Sepsis/Dehydration/DKA, Autoimmune / SLE, HUS
Trauma/ **T**umor : Trauma, Leukemia, lymphoma, SOL
Infections : Encephalitis / Meningitis, TBM, Brain Abscess
Cardiovascular: Cyanotic CHD, MS, AS, IE, Moya Moya, AVM, Vasculitis
Hematological: ITP, IDA, SCA, Protein C & S deficiency
Metabolic: Homocystinuria, Dyslipidemias

INVESTIGATIONS

a) Tabulated Overview

I. FOR DIAGNOSIS	II. TO ASCERTAIN CAUSE
1) Non- contrast Head CT	1) Blood: haematology
2) MRI Brain	2) Blood: biochemistry
3) MR/CT angiography	3) Urine: biochemistry
4) MR/CT venography	4) Blood immunology and infection screen
	5) Imaging

b) Details

I. FOR DIAGNOSIS (*Clinical and radiographic diagnosis*)

1) **Non- contrast Head CT imaging** (Acute HS & mature AIS)

2) **MRI** (For early & small infarcts) **Diffusion-weighted MRI **** (Demonstrates AIS within minutes of onset & up to 7 days post onset, + For wallerian degeneration in the descending corticospinal tract = chronic hemiparesis)

3) **MR/CT angiography cerebral and neck vessels** (for vascular occlusion and arteriopathy)
4) **MR/CT venography** (for CSVT : imaging of the cerebral venous system)

Tongue deviation	12th CN
Asymmetry of shoulder shrugging	11th
Asymmetry of palate elevation	9th
Eyebrow raising, tight closing of eyes, showing teeth and puffing out cheeks	7th
External eye movements	3rd ,4th ,6th
Visual fields *(A field defect implies a lesion at or above the internal capsule)*	2nd

3) **Examine the higher centres for parietal lobe signs**

Ask the child: '*Ankhain buund krain*'
Give pen in hand and ask: '*Yeah kia hai?*' (For Astereognosis)
Ankhain khol lain
Iss pen say farzi brush kar k dikhaain! (For ideomotor apraxia)
Pen say apna naam likhain (For Agrahia)
Circle buna k dikhain (For constructional apraxia: checks non-dominant side)

STEP V: GPE TO LOOK FOR A CAUSE

Hands	Clubbing,Cyanosis (cyanotic CHD), splinter hemorrhages (SBE), Pallor (SCA)
Pulse	Radioradial, radiofemoral delay
BCG scar	To Rule out TBM
Lymph nodes	To rule out Leukemia/Lymphoma
Scalp	Size: ?FOC (Subdural haematoma/intracranial tumour) Scars (e.g. craniectomy for AVM repair, evacuation of subdural haematoma) Sutures and fontanelles (wide sutures, full fontanelle with intracranial tumour, hydrocephalus) Shunts (e.g. hydrocephalus, chronic subdural collection) Auscultate the skull for bruits (AVM)
Eye	Conjunctivae for pallor (e.g. SCA), Leish nodules (NF), Cataract
Oral cavity	Inspect for haemorrhage from oral trauma
CVS	Murmurs (CHD, SBE, SLE), carotid pulsation (? in arteritis), carotid bruits
Abdomen	Hepatomegaly (SCA, ALL) and splenomegaly (SCA, SBE)
Feet	Edema

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) B.P. (to Rule out hypertension)(if Not done)
2) Urinalysis [For blood (e.g. SBE, post-streptococcal GN, SCA)][For protein (CKD)]
3) Fundoscopy [For retinal haemorrhage (NAI), papilloedema (raised ICP) or Roth spots (SBE)]
4) Vitals (missed ones)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place. He has **decreased movements of right side of body** with obvious **facial asymmetry**. There is no **dysmorphism, neurocutaneous stigmata** or **abnormal movements.**

HEMIPLEGIA/STROKE

1) General Look
2) Gait + Back
3) Motor system
4) Demonstrating the level of lesion
5) GPE to look for cause

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For dysphasia/intellectual impairment)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **P**osition patient [Lying/Sitting on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Note posture, asymmetry of the limbs (growth arrest)
2) Growth parameters (Tall/Marfanoid habitus, think of homocystinuria)
3) Well or unwell (e.g. encephalitis, SBE, CHD with cardiac failure)
4) Cyanosis (CHD)
5) Examine the skin

Bruising/purpura (e.g. non-accidental injury, Leukemia)	Pallor (e.g. Sickle cell anemia, Leukemia)
Neurocutaneous stigmata (e.g. S–W syndrome, NF-1)	

STEP II: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*
1) Romberg's sign
2) Full gait examination (esp Fog test and 'reverse Fog' test)
3) Back

Scoliosis (e.g. NF-1, spinal tumour)	Scars (e.g. excised spinal tumour)
Palpate for tenderness and masses	Auscultate ? (AVM, vascular tumours)

STEP III: MOTOR SYSTEM

See Motor system exam for details

STEP IV: DEMONSTRATING THE LEVEL OF LESION

1) **Inspect and describe facial features** (obvious facial asymmetry?)
2) **Examine motor cranial nerves** (12^{th} upward)

Social worker, occupational therapist, physiotherapist, speech therapist and psychologist

General	1. General nursing care (ALP – OF - TTNM CARE) 2. Pain 3. Schooling 4. Social implications
CNS	5. Seizures 6. Cognition/learning and communication difficulties 7. Sleep problems
Eye	8. Visual impairment
Ears	9. Hearing impairment
Mouth	10. Excessive salivation 11. Dental issues
Chest	12. Respiratory problems
Abdomen	13. GIT problems: dysphagia and nutritional issues
Limbs	14. Splints/orthoses 15. Management of spasticity

PROGNOSIS & COUNSELLING

Walking

Hemiplegic or diplegic CP	usually walk
quadriplegic CP	rarely walk
dyskinetic subtype	more difficult to predict
Sitting independently by 2 years	Most will walk
children who cannot sit by 4 years	unlikely to walk

Life span :

Only the most profound degrees of CP are associated with a decreased life expectancy.

If a child cannot lift the head to prone, and requires tube feeding, the median survival is around 17 years.

TIPS & TRICKS

1) Be gentle in handling CP child
2) Don't inflict pain
3) Be opportunistic in examination if you think that you will not be able to gain full cooperation

NOTES

An **IV cannula** on __ hand and **NG** in ___nostril are in place. He is wearing **nappies** but no **peripheral aids**.
He is **unable to walk**/has **Circumductive/scissoring gait**
Spine is normal
Muscle bulk is decreased with no **fasciculations**
Tone is increased in all limbs with fixed **contractures** at _____joints
Ankle Clonus is illicitable
Power is ___ in LL and ___ in UL
Deep tendon reflexes are exaggerated in all limbs
Plantars are upgoing/equivocal
Abdominal reflexes are intact and **bladder** is not palpable
Accessible motor cranial nerves are intact/ CN 7, 3,4,6 are intact
FOC is _____cm (appears decreased for age but I would like to plot it on centile charts)
Signs of meningeal irritation are absent
There are no signs of **pallor, clubbing, malnutrition, cataract, ear discharge, aspiration, murmur, hepatosplenomegaly, palpable stool pellets, bed sores** or **edema**.
BCG scar is present.
His **developmental age** appears to be _____months/years.

DIFFERENTIALS

1) Cerebral palsy
2) Post Meningitic/Kernicteric Sequele
3) Degenerative brain disease
4) Congenital Brain malformation

INVESTIGATIONS

I. FOR DIAGNOSIS	II. TO FIND ETIOLOGY	III. EXCLUSION OF OTHER DIAGNOSES
Clinical diagnosis	1) MRI Brain 2) TORCH screen 3) Urinary metabolic screen 4) Lumbar puncture	1) Thyroid function tests 2) Lactate, Pyruvate 3) ABGs & Anion gap (IEM) 4) S Ammonia (IEM) 5) Organic and amino acids 6) Chromosomal analysis 7) 24-hour day-cycle observation/ CSF for biopterin and neopterin 8) EEG

IV. TO RULE OUT COMPLICATIONS

1) Chest X ray
2) CBC
3) Urine C/S

MANAGEMENT & REHABILITATION

Multidisciplinary team approach
General paediatrician , orthopaedic surgeon, Eye speciatlist, ENT specialist

STEP IV - ROC

Three times (if time permits)

STEP V : CPE & RELEVANT

1) Signs of malnutrition
2) Hands. Pallor, clubbing
3) BCG scar
4) Lymph nodes
5) Eyes (Cataract, Vision, Nystagmus, Squint)
6) Hearing (look for any discharge)
7) Chest for aspiration
8) CVS for murmur
9) Abdomen for hepatosplenomegaly, Stool pellets
10) Genitalia (seek permission)
11) Back and buttocks (Scoliosis, Bed sores, sacral edema)
12) Edema
13) Hips for dislocation

STEP VI: DEVELOPMENT ASSESSMENT

Gross motor "180° manoeuvre", incorporating primitive reflexes:

1) Lying supine (assess position adopted, e.g. ATNR)
2) Pull to sit by hands (to assess head lag and grasp)
3) Sitting (assess sitting ability, then lateral propping)
4) Hold up vertically, under axillae (to detect increased extensor tone, scissoring, and automatic walking)
5) Tilt sideways (to assess head righting)
6) Ventral suspension (to detect ex passive extensor tone)
7) Parachute reflex (to detect asymmetry)
8) Place prone (to detect back arching)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS LONG CASE)*

1) Developmental assessment (if not done)
2) Fundoscopy
3) Vitals
4) Anthropometry
5) Primitive reflexes
6) Functional assessment (offer cup, spoon)
7) Formal visual & hearing assessment

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child who is **conscious** but I **could not gain full cooperation**. He has **flexed posture** with/without **abnormal movements**.
There is no **neurocutaneous stigmata**. **Bulbar** or **eye signs**.

CEREBRAL PALSY

1) General Look
2) Gait
3) Motor system exam
4) FOC
5) GPE & Relevant
6) Developmental assessment

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building
   One Important Question:
   Beta apka naam kia hai? (For hoarseness/slurring/Mental age)
c. **P**osition patient [Make him sit/lie on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

STEP I: INSPECTION

1) **Dysmorphic features** (e.g. chromosomal anomalies)
2) **Posture** (e.g. fisting, increased extensor tone, asymmetric tonic neck reflex , hemiplegic, quadriplegic)
3) **Movement**:
(a) Involuntary (e.g. choreoathetoid movements, dystonic spasms, seizures)
(b) Voluntary (e.g. immature gait pattern with wide base, up on toes, arms out for balance; hemiplegic, diplegic gaits; note posturing of arms when walking)
4) **Asymmetry** (e.g. hemiatrophy: look at the size of the thumbnails and the great toenails for subtle clues to asymmetry).
5) **Behaviour** (e.g. lack of interaction with environment, crying).
6) **Eye signs** (e.g. squint, nystagmus).
7) **Bulbar signs** (e.g. dysarthria, drooling)
8) **Interventions** (e.g. nasogastric tube, gastrostomy tube, scars of orthopaedic procedures).
9) **Clothing** (e.g. nappies in child over 4 years old).
10) **Peripheral aids** (e.g. wheelchair, splints, orthoses)
11) **Neurocutaneous lesions**

STEP II: GAIT

Ask: *'Bachaa Chal saktaa hai?'*(Proceed if Yes: Otherwise go to Step III)

STEP III: MOTOR SYSTEM EXAM

See Motor system exam for details

TREATMENT

After establishing my diagnosis by detailed history and clinical exam:

1) Multidisciplinary team approach
2) Counseling of Parents
3) Treatment of acute problem (Severe chorea: give diazepam, valproic acid , haloperidol , phenobarbitone)
4) Treat underlying cause

For Sydenham chorea	Benzathine pencillin prophylaxis
For Wilson's disease	Dietary restriction of nuts, chocolates, Pencillamine with zinc acetate
For SLE	Steroids, Hydroxychloroquine

PROGNOSIS

Depends on cause

1) Self limiting (may take 3-6 months)
2) 20% cases recur

TIPS & TRICKS

1) If command is motor system examination then do motor system first and then GPE & relevant systemic exam.

NOTES

STEP VI: MOTOR SYSTEM & CN

1) Motor system (Lower limb first then upper limb)
2) Cranial nerve 7

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) BP
2) Anthropometry
3) Functional assessment
4) Slit lamp exam for KF rings

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious & cooperative** with an IV cannula in place.
There is no **dysmorphism, deformity, neurocutaneous stigmata.**
He has **Choreform movements involving**_____(body parts involved), **Un-intangible Speech** and **emotional lability** with no **distress.**
He is showing **Signs of Chorea** in form of spooning of hands, Milk maid sign, Pronator sign and Darting tongue.
Gait is _____ and **spine** is normal.
He is **afebrile** and there is no evidence of

a. **Subcutaneous nodules, erythema, joint swelling** (Rheumatic fever)
b. **Clubbing, Hepatosplenomegaly, Ascites, Jaundice** (CLD)
c. **Oral ulcers, rash, alopecia** (SLE)

Apex beat is un displaced with normal character. Both heart sounds are audible and there is no murmur.
Muscle bulk is normal and symmetrical in all limbs with no scar marks or obvious deformity.
Tone is decreased/normal, **power** grade 5/5 with normal **DTR** and **plantars**.

DIFFERENTIALS

1) Sydenham chorea (Rheumatic fever)
2) Wilson's disease
3) SLE
4) Drug induced (L-Dopa, Phenytoin, Valproate, Carbamazepine, TCAs, Phenothiazines)
5) Cerebral palsy

INVESTIGATIONS

For Sydenham chorea	CBC , ESR, CRP, ECG, CXR, ECHO
For Wilson's disease	LFTs, Serum ceruloplasmin level, 24 hr urinary copper level, Slit lamp examination for KF rings, USG Abdomen
For SLE	ANA, Anti DsDNA, anti phospholipid antibody level
For Drug induced	Drug levels
For CP	MRI Brain

CHOREA

1) General Look
2) Signs of Chorea
3) Gait & Back
4) GPE
5) CVS, Abdomen
6) Motor system (LL then UL)

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For hoarseness/slurring)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **P**osition patient [Make him sit on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

1) Respiratory rate (at foot end)
2) Look for dysmorphism, rash, Jaundice

STEP II: SIGNS OF CHOREA

1) **Spooning of hands** (On projection of hands forward, flexion at wrist, extension of finger joints & abduction of thumb)
2) **Milk maid sign/grip** (Ask patient to grip your hands; grip increases and decreases like milking)
3) **Pronator sign** (On raising arms above head; Pronation of one or both arms)
4) **Darting tongue/Jack in the box tongue** (Ask to protrude tongue out: Protrudes in & out)

STEP III: GAIT & BACK

Ask: *'Bachaa Chal saktaa hai?'*

STEP IV: MAKE HIM SIT ON COUCH FOR GPE

1) Fever, Pulse, Subcutaneous nodules, erythema marginatum, joint swelling (Rheumatic fever)
2) Signs of CLD, Pallor, Jaundice (CLD)
3) Oral ulcers, rash, alopecia (SLE)

STEP V: ASK TO LIE ON COUCH FOR CVS & ABDOMEN

1) CVS: Murmur for RHD (MR, AR)
2) Abdomen: Liver, spleen, ascites (CLD, SLE)

INVESTIGATIONS

1	**For Infectious causes**	CBC, CRP, Viral serology, CSF analysis
2	**For Drug induced**	Drug levels
3	**For SOL/DBD**	MRI Brain
4	**For Ataxia Telangiectasia**	Chromosomal breakage studies Serum immunoglobulin levels (Decreased IgG,IgA but Increased IgE) Increased alpha feto protein level
5	**For Friedrich's Ataxia**	Electrophysiologic studies: Abnormal visual, auditory brainstem, and somatosensory-evoked potentials ECG, CXR, 2D Echo (For cardiomyopathy) X-ray spine (For Khyphosis/scoliosis)
6	**For Abetalipoproteinemia**	Peripheral blood smear (for Acanthocytes) Decreased triglyceride & cholesterol Absent beta lipoproteins Decreased Vit E levels

TREATMENT

1) Multidisciplinary team approach
2) Counseling of Parents
3) Supportive treatment
4) Treat underlying cause

PROGNOSIS

Depends on cause

1) Acute (Post viral) : Self limiting
2) SOL : Good after surgery
3) DBD: Poor

TIPS & TRICKS

1) Skip test for Rebound phenomenon as it may hurt the child.
2) Compare coordination of both right and left side
3) Enumerate both acute and chronic causes in short case as history is not available.
4) Labrynthitis can be ruled out due to obvious cerebellar signs.
5) Lesions affecting Vermis (Central part) of cerebellum ? Truncal Ataxia
6) Lesions affecting hemispheres of cerebellum ? Appendicular (Limb) Ataxia
7) Lesions affecting Flocculonodular lobe (Below vermis) of cerebellum ? Posture instability, Impaired eye movement control

NOTES

STEP V: MOTOR SYSTEM EXAMINATION

See Motor system exam for details

STEP VI: RELEVANT GPE

1) **Locate apex beat and auscultate** (Cardiomyopathy in Friedrich's Ataxia)
2) Look for **Ear discharge**

STEP VII: ASK TO GET UP WITH ARMS CROSSED

For Truncal Ataxia

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Proprioception
2) Fundoscopy (SOL)
3) Vitals
4) Anthropometry
5) Otoscopy (For Ear pathology)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious & cooperative** with an IV cannula in place.
He/She has slurred **(dysarthric) speech** and **ataxic gait** with **inability to perform Tandem walking**.
Romberg is positive/negative
Spine is normal/There is evidence of Kyphosis/Scoliosis
I have appreciated Signs of Cerebellar lesion in form of:

Horizontal/Vertical nystagmus	**Intentional tremors**	**Past pointing**
Dysdiadokokinesia	**Pendular knee jerk**	**Poor heel shin coordination**
And **Truncal Ataxia**		

He/she is **hypotonic** but **Muscle bulk, power, DTR and plantars** are normal
There is no evidence of

Pallor	Jaundice	Telangiectasia	Ear discharge
Petechiae, Bruise, Rash		Pes cavus	Hammer toes

Ataxia: 'Loss of control of body movements'

DIFFERENTIALS

Acute Cerebellar ataxia	Chronic Cerebellar Ataxia
1) Post infectious (Varicella, Coxsackie, Echovirus) 2) Drug induced (Phenytoin, Thallium, Alcohol) 3) Traumatic	1) Cerebellar Abscess 2) Tumor (Neuroblastoma) 3) Degenerative brain diseases (Ataxia Telangiectasia, Friedrich's Ataxia) 4) Abetalipoproteinemia 5) Congenital Anomalies of Posterior Fossa (Dandy walker malformation, Arnold Chiari Malformation, Agenesis of cerebellar Vermis)

CEREBELLAR EXAM/ATAXIA

1) General Look
2) Gait & Back
3) Cerebellar signs (Eye, Finger-Nose ,Hands, Knee, Heel-Shin)
4) Motor System
5) Relevant GPE
6) Testing for Truncal ataxia

WIPER

a. Wash your hands with sterilizing solution/Warm your hands
b. Introduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For hoarseness/slurring)
   2) *Kis class main parhtay hain?* (For Mental age)
c. Position patient [Make him sit on couch/parent's lap]
d. Exposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. Right side to approach for examination

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

Petechiae, Bruise, Rash (Varicella)
Telangiectasia on nose, upper chest (Ataxia Telangiectasia)
Ask: *'Bachaa Chal saktaa hai?'*(Proceed if Yes: Otherwise go to Step III)

STEP II: GAIT & BACK

1) **Gait maneuvers** (Wide based ataxic gait) + Heel-toe walking (Unable to perform)
2) **Romberg test**
3) **Check Back** for Scoliosis/Kyphosis

STEP III: MAKE CHILD SIT ON COUCH FOR CEREBELLAR SIGNS

1) **Eye examination**

Horizontal & Vertical Nystagmus	Pallor (Vit B12 deficiency)	Jaundice (CLD/Abetalipoproteinemia)	Telangiectasia

2) **Extending upper limb**: note any drift or static tremor (due to hypotonia)
3) **Finger-Nose test (Intentional tremors**, Past pointing/**Dysmetria**)
3) **Rapidly pronate and supinate the hand** (for **dysdiadochokinesis**)
4) **Pendular Knee jerk**

STEP IV: MAKE CHILD LIE ON COUCH

1) **Heel Shin test** (Coordination of Lower limbs)
2) **Check for Pes cavus/Hammer toes** (Friedrich's Ataxia)

5) Bladder & bowel care 6) Physiotherapy	5 days) **OR** (1g/kg for 2 days) 5) Plasmapheresis(250ml/kg in 4-6 session) 6) Steroids (CIDP/Relapse)	3) Physiotherapy	

TIPS & TRICKS

1) Avoid inflicting pain to child. Handle gently.

NOTES

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Vitals (if Not done)
2) Sensory system examination (if not done)
3) Fundoscopy
4) Anal tone

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
There is no **dysmorphism, deformity, neurocutaneous stigmata or abnormal movements**
Spine is normal
There is no **hoarseness** and **respiratory distress**.(Respiratory rate = ___/min)
He is moving his arms but not legs **(Posture)**
He is unable to walk. **(Gait)**
Muscle bulk is normal & symmetrical in all limbs. There are no scar marks or obvious deformity. **(Inspection of limbs)**
He has sign of lower motor neuron lesion in lower limbs in form of hypotonia, power of grade 2/5 , absent DTR & flexor plantar reflex. **(Tone Power Reflexes)**
Superficial abdominal ± cremesteric reflexes are present. **(Superficial reflexes)**
Abdomen is soft and bladder not palpable. **(Abdomen)**
Motor exam of upper limb & cranial nerves is normal. **(UL & CN)**
BCG scar is present. **Pulse** is 80/min, regular in rhythm & normal in volume & character.

DIFFERENTIALS

Symmetric	Asymmetric
1. Peripheral neuropathy (GBS) 2.Myelopathy (Transverse myelitis)	1. Polio 2. Traumatic neuritis

INVESTIGATIONS

POLIO	GBS	T.MYELITIS	T.NEURITIS
2 stool cultures 24-48 hrs apart	EMG/NCS CSF Gadolinium enhanced MRI of spinal cord Antigangliosidase Ab	MRI spine	NCS

TREATMENT

1)**Multidisciplinary team approach**
2) **Inform WHO AFP team**

POLIO	GBS	T.MYELITIS	T.NEURITIS
1) Proper positioning 2) Analgesics 3) Sedatives 4) Hydration and diet	1) Analgesics 2) Ventilatory support 3) Physiotherapy 4) IVIG (400mg/kg/day for	1) High dose Methyl prednisolone 2) Bladder catheterization	1) Analgesics

ACUTE FLACCID PARALYSIS

1) General Look
2) Gait
3) Motor system & Abdomen
4) Cranial nerves
5) Vitals, CVS, Chest & Back
6) Sensory system

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For hoarseness/slurring)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **Position patient [Lying on couch/parent's lap]**
d. **Exposure:** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: GENERAL LOOK

General Look; vitals (Respiratory Rate), Posture

STEP II: GAIT

Ask: *'Bachaa Chal saktaa hai?'*
Check Gait (mostly unable to walk)

STEP III: MOTOR SYSTEM & ABDOMEN

See Motor system exam for details
Palpate abdomen for Bladder & impacted feces

STEP IV: CRANIAL NERVES

Ask to cough; CN 3, 6, 7 (for miller fisher variant)
'AAh test' for uvula (CN 9,10) (Bulbar involvement)

STEP V: VITALS, CVS, CHEST & BACK

Vitals: Pulse rate, B.P.
CVS: Arrythmias
Chest: Lung bases
Back: Spine, MMC, Scar, Bed Sores, Mass

STEP VI: SENSORY SYSTEM

Redress the child and say thank you!

Scissor like gait (Spastic diplegia)	Shuffling gait (Parkinsonism)

2) **Heel–toe walking**: (>2yr old)

Inability to perform Heel-toe walking		
Pathology in cerebellar vermis	Weakness	Sensory deficits

3) **Walking on toes**: Tests strength of plantar flexion (S1)

Usually possible to Perform	Inability to perform (lesions affecting S1)
Cerebral palsy Duchenne muscular dystrophy	Low lumbar myelomeningocoele Peripheral neuropathies Anterior horn cell disease

4) **Walking on heels**: Tests strength of dorsiflexion (L5)/contractures of the calf muscles

Inability to perform Heel walking			
CP	DMD	Anterior horn cell disease	Peripheral neuropathies

5) **Walking on outsides of feet** (Fog test): Asymmetry in arm and leg positioning (Hemiplegia)
6) **Walking on the insides of feet** (Reverse Fog): Similar significance as of Fog test

STEP IV: RUNNING

Accentuates findings such as **hemiplegia** and **proximal weakness** (in the latter, a child may seem to be miming a run in slow motion)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Motor system exam (esp. Lower limb)
2) Fundoscopy (for SOL)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
There is no **dysmorphism, deformity, neurocutaneous stigmata or abnormal movements**
Spine is normal
Romberg is positive/negative
Gower's Sign is positive/negative
He/ She has **Normal/Antalgic/Circumductive/Waddling gait**
He/ She is/isn't able to perform **Heel-toe walking, Toe walking, Heel walking, Fog test** and **Reverse Fog test**
He/She is able/unable to run normally

DIFFERENTIALS

1) Neurological problem
2) Orthopedic problem
3) Rheumatological problem
4) Hemophilia

GAIT EXAMINATION

1) Inspection
2) Standing (Romberg, Trendelenberg, Hop, Gower, Spine)
3) Walking (Normal, Heel toe, On toes, On heels, Fog, Reverse fog)
4) Running

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For hoarseness/slurring)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **Position patient** [Standing on both feet with attendant nearby]
d. **Exposure:** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

STEPS OF EXAMINATION

STEP I: INSPECTION

1) **Head to toe examination**

Large head	Eye signs (squint, ptosis, nystagmus)
Abnormal posture (Hemiplegia)	Abnormal movements (Tremors, fasciculations)
Asymmetry (Growth arrest with hemiplegia, Rib asymmetry with scoliosis)	
Neurocutaneous stigmata	Scars of procedures (e.g. tendon releases, VP shunts)

2) **Examination of Limbs** [Muscle bulk, wasting, contractures and any deformities of the feet (e.g. talipes, pes cavus), Calf hypertrophy & Thigh muscle wasting (DMD)]

STEP II: STANDING MANOEUVRES

1) **Standing with feet together** (Romberg test)

Swaying/falling With eyes open	Swaying/falling With eyes closed
Truncal (cerebellar) ataxia	Romberg's sign +ive (Dorsal column pathology)

2) **Standing on each foot** (Trendelenberg's sign): Inspect from back for position of pelvis, by iliac crest position (It detects any proximal instability and positive Trendelenberg's sign)
3) **Hopping on each foot:** (For unilateral weakness and for balance)
4) **Squatting and then rising/ Lying on the floor and then rising** (Gower's manoeuvre)

Difficulty maintaining squat	Positive Gower's sign
Peripheral neuropathy	Proximal weakness (DMD)

5) **Bending forward and touching toes** : (for scoliosis, hairy patches, MMC scars, lipomas)

STEP III: WALKING MANOEUVRES

1) **Normal gait**: focus on the pelvis, hips, knees and feet

Circumductive (Hemiplegia)	Wide-based (Cerebellar dysfunction)
Waddling (Proximal myopathies)	Antalgic gait (limp) (Orthopaedic problems)

1) To check power in unconscious/child Not obeying command: Stroke sole of feet with finger and check movement.
2) While checking deep tendon reflexes, if reflex is not elicited; try twice and then try with reinforcement. If still reflex is not elicited, move to the other limb.
3) Remember dermatomes.
4) Skip contradictory findings.

NOTES

2) **Palpate bladder**

STEP IV: 'TPR' UPPER LIMB

Check **Tone, Power & Reflexes** of upper limb

STEP V: ACCESIBLE MOTOR CRANIAL NERVES

1) Cranial nerves 7
2) Make H to check Eye movements (CN 3,4,6)

STEP VI: HEAD & NECK

1) FOC
2) Check for neck rigidity

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Rest of the Cranial nerves (if Not done)
2) Cerebellar exam (if relevant)
3) Sensory system (if relevant)
4) Developmental assessment (if relevant)
5) Fundoscopy (if relevant)
6) Vitals

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
There is no **dysmorphism, deformity, neurocutaneous stigmata or abnormal movements**
Spine is normal
Romberg is positive/negative
He/ She has **Normal/Antalgic/Circumductive/Waddling gait**
Muscle bulk is normal with no **fasciculations**
Tone is Normal/Increased/decreased in all limbs
Ankle Clonus is/isn't illicitable
Power is 2/5 in LL and 5/5 in UL
Deep tendon reflexes are Normal/exaggerated/absent in all limbs
Plantars are downgoing/upgoing/equivocal
Abdominal reflexes are intact and **bladder** is not palpable
Accessible motor cranial nerves are intact/ CN 7, 3,4,6 are intact
FOC is _____cm (appears normal/decreased for age but I would like to plot it on centile charts)
Signs of meningeal irritation are absent

TIPS & TRICKS

1) In motor system doing Gait first can been the best screening method to reach a diagnosis.
2) While checking gait properly expose the lower limbs and make sure someone is beside child to prevent fall.
3) Check power in both limbs simultaneously to save time.

MOTOR SYSTEM

1) General Look
2) Gait & Rapid Motor scan
3) Tone,Power, Reflexes (TPR)
4) Abdominal reflexes & Bladder
5) Cranial nerves
6) FOC, SOMI

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For hoarseness/slurring)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **P**osition patient [Make him sit on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

Ask: *'Bachaa Chal saktaa hai?'*(Proceed if Yes: Otherwise go to Step II)

STEP I: MAKE CHILD STAND FOR 'RAPID MOTOR SCAN'

1) **Inspection from front** (dysmorphism, deformities, neurocutaneous lesions, involuntary movements, Fasciculations, wasting, hypertrophy)
2) **Inspection from back** (calf, asymmetry, buttocks, spine) (Ask to bend & check spine)
3) **Romberg test**
4) **Ask to bring hands in front & do sit stand/sit up** (to scan power of all muscles) (Illicit complete Gower sign if suspecting DMD)
5) **Gait** (simple walk, turning around, tandem walking) Ask attendant to be near child

STEP II: LIE DOWN ON COUCH FOR 'TPR' LOWER LIMB

'Beta! Kahiin dard tuu nahi hai'
1) **Palpate muscles** (look into eyes of child)
'Hath paoon bilkul dheelay chor dain'
2) **Tone & Clonus** assessment (while talking to child) (Passive movements at major joints: Roll? flex knee? movement at ankle? check ankle clonus)
3) **Power** (Hip: Flexion,extension, adduction, abduction) (Ankle: Dorsi flexion, Plantar flexion, eversion, inversion) (Knee: Flexion, extension)
4) **Deep tendon reflexes** (Knee, Ankle)
5) **Superficial reflexes** (Plantars: with car key)

STEP III: ABDOMEN

1) **Superficial reflexes** (Abdominal reflexes with car key)

MOTOR SYSTEM

1) General Look
2) Gait & Rapid Motor scan
3) Tone,Power, Reflexes (TPR)
4) Abdominal reflexes & Bladder
5) Cranial nerves
6) FOC, SOMI

WIPER

a. **W**ash your hands with sterilizing solution/**W**arm your hands
b. **I**ntroduction & Rapport building
   Two Important Questions:
   1) *Beta apka naam kia hai?* (For hoarseness/slurring)
   2) *Kis class main parhtay hain?* (For Mental age)
c. **P**osition patient [Make him sit on couch/parent's lap]
d. **E**xposure: (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

Ask: *'Bachaa Chal saktaa hai?'* (Proceed if Yes: Otherwise go to Step II)

STEP I: MAKE CHILD STAND FOR 'RAPID MOTOR SCAN'

1) **Inspection from front** (dysmorphism, deformities, neurocutaneous lesions, involuntary movements, Fasciculations, wasting, hypertrophy)
2) **Inspection from back** (calf, asymmetry, buttocks, spine) (Ask to bend & check spine)
3) **Romberg test**
4) **Ask to bring hands in front & do sit stand/sit up** (to scan power of all muscles) (Illicit complete Gower sign if suspecting DMD)
5) **Gait** (simple walk, turning around, tandem walking) Ask attendant to be near child

STEP II: LIE DOWN ON COUCH FOR 'TPR' LOWER LIMB

'Beta! Kahiin dard tuu nahi hai'
1) **Palpate muscles** (look into eyes of child)
'Hath paoon bilkul dheelay chor dain'
2) **Tone & Clonus** assessment (while talking to child) (Passive movements at major joints: Roll? flex knee? movement at ankle? check ankle clonus)
3) **Power** (Hip: Flexion,extension, adduction, abduction) (Ankle: Dorsi flexion, Plantar flexion, eversion, inversion) (Knee: Flexion, extension)
4) **Deep tendon reflexes** (Knee, Ankle)
5) **Superficial reflexes** (Plantars: with car key)

STEP III: ABDOMEN

1) **Superficial reflexes** (Abdominal reflexes with car key)

Pulsations

1) Aneurysm
2) Swelling over major blood vessel
3) Highly vascular tumor

Bruit

1) Aneurysm
2) AV malformation
3) Highly vascular lump

Tender	Non-tender
Boil	Lipoma
Abscess	NF
Lymphadenitis	Tuberculous lesions

Erythematous	Warm
Inflammatory	Inflammatory
Hematoma	Rapidly growing

NOTES

a. Move lump in different planes relative to surrounding tissue (to assess f ixation with deeper structures)
b. Fluctuation in two planes (for fluid / soft lipomas) compress on one side; see if a bulge occurs on the other side

II) AUSCULTATION

For Vascular bruits

III) RELEVANT

a. Other sites of similar lumps / swellings (DD: NF, Lipomatosis, Lymphomas)
b. Assess draining lymph nodes

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Transillumination (if Not done)
2) Vitals (if relevant)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff)
_____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place.
A **6 × 8cm spherical** swelling is noted in **Right cervical region**
Overlying skin is → Erythematous, warm & of normal texture with/without discharge/sinus
→ of normal color, temperature & texture with/without discharge/sinus
Swelling is **non-tender/ tender** with **sharp / rounded margins, smooth / irregular surface & soft / firm / hard consistency**.
It is **non-mobile / mobile & attached** to Skin/Underlying Structures
No **pulsations / thrill or bruit** is noted.

DIFFERENTIALS

Very Hard consistency	Fluctuation present
1) Storage	1) Blister
2) Malignancy	2) Abscess
3) Calcified	3) Cyst
4) Dense Fibrous tissue	4) Lipoma
Surface	**Margins**
1) Smooth (e.g. AVH)	1) Regular (Enlarged organ e.g. thyroid)
2) Irregular (e.g. CLD, Metastasis)	2) Irregular (Inflammatory/Malignant)

EXAMINATION OF LUMP/SWELLING/LYMPH NODE

1) Inspection
2) Palpation
3) Movements
4) Auscultation
5) Relevant
6) Transillumination

WIPER

a. **Wash your hands with sterilizing solution/Warm your hands**
b. **Introduction & Rapport building**
c. **Position patient** [Make him sit/lie on couch depending upon location of lump]
d. **Exposure: EXPOSE, EXPOSE, EXPOSE** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **Right side to approach for examination**

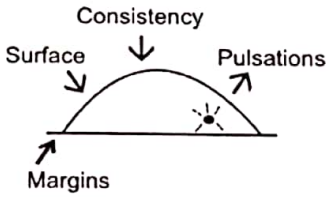
STEPS OF EXAMINATION

I) INSPECTION (5 × S)

a. **Site**
b. **Size** (Measure with inch tape)
c. **Shape**
d. **Skin overlying** [Lymphatic obstruction → Skin Fixation & fine dimpling at opening of hair follicles = Peaud' orange (orange peel)]
e. **Sinus/Discharge**

II) PALPATION *(Ask: 'Iss sojaan main dard tuu nahi?')*

a. Gentle palpation for tenderness (Look for signs of distress on face)
b. Compare temperature with surrounding skin (Dorsum of Hand)



c. Margins / Edge (Regular/Irregular; sharp/rounded)
d. Surface texture (Smooth/Irregular e.g. Nodular)
e. Consistency (Soft, Firm, Hard, Stony Hard)
f. Pulsatile / non-pulsatile (feel it) (Time with Radial Pulse) + Thrill

III) MOVEMENTS

a. Pick up overlying skin fold (to assess attachment with Skin)

Positive examination findings: _____
Important negative examination findings: _____

TIPS & TRICKS

1) Usually the most difficult cases are when command is: Do GPE and relevant.
2) Examiners want you to reach diagnosis by performing GPE. Focusing on any specific system without completing GPE can lead to failure.
3) If you find nothing on general look then make the child stand, do anthropometry first.
4) Hidden cases in GPE : ITP (petechiae), Hemophilia (Joint swelling), Leukemia/Lymphoma (Lymph nodes, Bruise) , Ambiguous genitalia/Precocious puberty
5) Do not spend so much time on hands and arms.
6) Four areas to cover in GPE are : 1) General Look, 2)Vitals, 3) Anthropometry, 4) Hand →Head →Toe → Back : At least touch each of them e.g. take 2-3 vitals/1-2 Anthropometric measurements (Offer missed ones)
7) Practice to check B.P. in 20 seconds (Essential in edematous/obese child, CVS cases, CKD, Abdominal mass, Ambiguous genitalia)
8) Never waste your time e.g. if you can see that there is no neck swallowing and child is not swallowing despite you asked him twice to swallow, move on.
9) Place thermometer in the left armpit because from the right arm you will check B.P. And never forget to remove thermometer from armpit.

NOTES

1. **Cervical & Supra clavicular Lymph nodes**
2. **Posterior Axillary lymph nodes**
3. Check **spine** (tenderness / deformity, sacral edema, MMC)

(POSITION = SUPPORTED AT 45 DEGREE WITH YOUR HAND)

1. Check **JVP** (using light of torch)

(POSITION = LYING FLAT ON COUCH) (General & Relevant exam)

'Chaati yaa paait main kahiin daard tuu nahi?'

2. **Chest** (Inspection), Check Sternal tenderness
3. **Abdomen** (Inspection), palpation (if relevant)
4. **Seek Permission for Genitalia** (SMR Staging), inguinal lymph nodes, Scrotal Edema (If girl: seek permission and do not proceed until permission is granted. If boy: Tell examiners and the patient and proceed) (**Can be a case of ambiguous genitalia/precocious puberty**)
5. Check **Knee** for Swelling, **Legs** for rash/bruises
6. Pedal **Edema**

(POSITION = TILTING TO A SIDE ON COUCH/STANDING)

1. **Buttocks** (For wasting, Rash, Bone Marrow Scar)

STEP III: VITALS

If missed previously

STEP IV: ANTHROPOMETRY

Ask : Bacha chal sakta hai?

1. Check **Gait**
2. **Anthropometry** (Height, weight, FOC)

Redress the child and say thank you!

OFFER *(DO IF TIME PERMITS/LONG CASE)*

1) Blood pressure (if Not done)
2) Fundoscopy (if relevant)
3) Otoscopy (if relevant)

DESCRIPTION

Thank you sir! I would like to complete my examination by doing _____ (missed stuff) _____(Name) ___ yrs old child who is **conscious** and **cooperative** with IV cannula in place. There are No **Dysmorphic** features, Signs of **D**ehydration, **J**aundice, **P**allor ,**C**yanosis or Resp distress. **(DYSMORPHIC DJ-PCR)**
The Child is interacting appropriately for his/her age. **(BEHAVIOUR)**
There are
-No **P**urpuric spots, **P**igmentation (hypo or hyper)
- any **R**ash
-or **S**cratch marks visible on Skin
Note: describe rash if found.
With RR_____, HR_____, B.P. _____, and Temperature (axillary) _____, Weighing _____ with Height/Length of ____ & Fronto-Occipital Circumference of _____(centiles). (I would like to plot them on centile charts)

GENERAL PHYSICAL EXAMINATION

1) General Look
2) Hand → Head → Toe → Back
3) Vitals
4) Anthropometry

WIPER

a. **W**ash your hands with sterilizing solution/Warm your hands
b. **I**ntroduction & Rapport building
c. **P**osition patient [Make him sit on couch/parent's lap]
d. **E**xposure: **EXPOSE, EXPOSE, EXPOSE** (Ideal exposure in male child: Shirt off, trousers rolled up) [Seek parent's help to undress] (Beware in case of elder girl)
e. **R**ight side to approach for examination

STEPS OF EXAMINATION

(Listen to command & Age of child)
(Ask yourself: **'Is his height appropriate for age**?' If short stature: Do anthropometry first)

STEP I: GENERAL LOOK

(POSITION = CHILD SITTING ON COUCH)

1. **LOOK, LOOK, LOOK** for a spot diagnosis. (e.g. Down's syndrome, Cushingoid features, Hydrocephalus, Pallor, Facial palsy, Jaundice Nephrotic, goiter, rickets, renal osteodystrophy, target joint, MPS, chorea, dystonia and myasthenia gravis)
2. **RR**: Stand at a distance & Count respiratory rate (for 6 sec → multiply by ten)

STEP II: HAND ? HEAD ? TOE ? BACK

(POSITION = CHILD SITTING ON COUCH)

1. **Hand shake** (ideally both hands) (Check temperature,sweaty,skin texture,grip)
2. **Hand**, Nails, Palm, Palpate finger pulp (check clubbing with card)
3. **Forearm**: Check for radius, widening of wrist, Mantoux test mark
4. **Pulse** rate & rhythm (for 6sec & multiply by 10)
5. Move hand along **extensor surface** (for rheumatic nodules)
6. **Epithrochlear nodes**
7. **BCG scar** mark (check on right Side → check on left if not found)
8. Check **BP** (In cases of Nephrotic, CKD, CVS) (Avoid arm with fistula)
9. **Temperature** with thermometer/back of hand
10. **Axillary Lymph nodes**
11. **Occipital & Post auricular Lymph nodes**
12. **Heads & Scalp** (Hair, Anterior Fontanelle)
13. **Eyes**
14. **Nose, ear**
15. **Oral cavity** & tongue
16. Neck → **Thyroid** (ask to swallow)

FROM BACK (Ask: *'Kamaar main kahiin daard tuu nahi?'*)

8. WEIGHT

New born	-	2.5-3.5 kg
6 months	-	7.5 kg
1 yr - 7 yr	(Age +4) x 2	
8 yr – 9 yr	(Age +5) x 2	
10 years	(Age +6) x 2	32 kg
11 years	(Age +7) x 2	36 kg
12 years	(Age +8) x 2	40 kg

5. FRONTO-OCCIPITAL CIRCUMFERENCE (FOC)

New born	**35cm**
3 months (Increases by 6cm)	41cm
6 months (Increases by 3cm)	44cm
9 months (Increases by 2cm)	46cm
1 year (Increases by 1cm)	**47cm** (Pak: 1947)

2 cm rise/yr

2 years	49cm

1 cm rise/yr

3 years	**50cm**

0.5 cm rise/yr

4 years	50.5cm
5 years	51cm
6 years	**51.5cm**
7 years	52cm
8 years	52.5cm
9 years	53cm
10 years	**53.5cm**
11 years	54cm
12 years	**54-55cm**

7.LENGTH/HEIGHT

New born	**50cm**
1 year	75cm (*25cm rise*)
2 years	85cm (*10cm rise*)
3 years	**95cm** (*10cm rise*)

95 + 6cm / year

4 years	101cm
5 years	107cm
6 years	**113cm**
7 years	119cm
8 years	125cm
9 years	131cm
10 years	**137cm**
11 years	143cm
12 years	**149cm**

AJ'S ART OF REMEMBERING VITALS

1. RESPIRATORY RATE (WHO IMCI guidelines)

Age group	Respiratory rate (Upper limit)	Heart Rate
0-2 months	60/min	
2-12 months	50/min	
1-5 yr	40/min	
6-12 yr	22/min*	
12 yr +	18/min*	

* Nelson 21ed

2. HEART RATE (2-3 times normal respiratory rate for age)

Age group	Respiratory rate (Upper limit)	Heart Rate
0-2 months	60/min	120-180/min
2-12 months	50/min	100-150/min
1-5 yr	40/min	80-120/min
6-12 yr	22/min	66/min
12 yr +	18/min	54/min

3. SYSTOLIC BLOOD PRESSURE

Age group	Lower limit	Upper limit
0-1 months	60 mm Hg	85 mm Hg
1-12 months	70 mm Hg	90 mm Hg
1-10 yr	70 mm Hg + (2 × age)	90 mm Hg + (2 × age)
10 yr +	90 mm Hg	120mm Hg

4. TEMPRATURE

(Place thermometer in armpit for 2 min)

Normal temperature 97.5^0 F – 99.5^0 F

NEPHROLOGY

CHRONIC KIDNEY DISEASE	EDEMA/NEPHROTIC SYNDROME
1) CKD 2) Celiac disease (short + pale) 3) Rickets (Bony deformities)	1) Nephrotic syndrome 2) Nephritic syndrome i.e AGN 3) To rule out CLD, CCF, Protein losing enteropathy

RESPIRATORY

CONSOLIDATION	PLEURAL EFFUSION
1) Pneumonia 2) Tuberculosis 3) Collapse with patent main bronchus	1) Parapneumonic 2) Post Tuberculosis 3) Empyema 4) Collapse with obstructed main bronchus

BRONCHIECTASIS

1) Post tuberculosis
2) Post pneumonic
3) Post viral (Pertussis, measles)
4) Cystic fibrosis
5) Primary ciliary dyskinesia
6) Immunodeficiency
7) Congenital lung lesion
8) Interstitial lung disease

MUSCULOSKELETAL/AUTOIMMUNE

DUCHENNE MUSCULAR DYSTROPHY	JUVENILE DERMATOMYOSITIS
1) Duchenne muscular dystrophy	1) JDM
2) Becker muscular dystrophy	2) Juvenile polymyositis
3) Limb girdle muscular dystrophy	3) SLE
4) SMA-3	4) MCTD

JUVENILE IDIOPATHIC ARTHRITIS

(*Mnemonic: Joints ache* ***In TrAM***)

Infections	Septic arthritis Osteomyelitis Reactive arthritis Tuberculous arthritis Acute rheumatic fever
Trauma	Local trauma Avascular necrosis (Legg-Calvé-Perthes disease) Slipped capital femoral epiphysis
Autoimmune	JIA SLE JDM/MCTD/Scleroderma IBD Autoimmune hepatitis
Malignancy & Hematology	Hemophilia Sickle cell disease Leukemia Neuroblastoma Bone tumor (osteosarcoma, Ewing sarcoma)

SYSTEMIC LUPUS ERYTHEMATOSUS

Infections	Sepsis, Epstein-Barr virus, parvovirus B19, endocarditis
GN	Poststreptococcal glomerulonephritis
Rheumatologic	sJIA, vasculitides
Malignancies	Leukemia , Lymphoma
Drugs	Antibiotics: Minocycline, Sulfonamides, tetracycline, penicillin

NUTRITION

MARASMUS	KAWASHIORKER
Primary malnutrition	Primary malnutrition
Celiac disease	Protein losing enteropathy
Chronic infections (T.B./HIV)	CLD
Chronic illnesses (CHD, CLD, CKD)	Nephrotic syndrome

RICKETS/BOWING OF LEGS

Younger child	Older child
Nutritional rickets Vit D Resistant rickets CKD	Vit D dependent rickets X-linked hypophosphatemic rickets CKD

HEMATOLOGY/ONCOLOGY

Groups	HSM+Pallor	HSM+Jaundice	HSM+Ascites
1.Infections	Malaria Disseminated T.B. Kala-azar IE	Hepatitis Malaria Disseminated T.B. (icterus is due to drugs)	Hepatitis Abdominal T.B.
2.Hematological	Thalassemia Sickle cell HS	Thalassemia Sickle cell HS	Thalassemia
3. Malignancy	Leukemia Lymphoma	Leukemia Lymphoma	Leukemia Lymphoma
4. Metabolic	Wilson Gaucher	Galactosemia Alpha-1 antitrypsin deficiency	Galactosemia Alpha-1 antitrypsin deficiency
5. Misc	SLE Cirrhosis with portal HTN	Drugs Cirrhosis with portal HTN	Drugs Cirrhosis with portal HTN CCF

Groups	HSM+Lymphadenopathy	HSM+Bleeding
1.Infections	Disseminated T.B. Kala-azar HIV	Septicemia TORCH infections Dengue IE
2.Hematological	Thalassemia	Thalassemia HSP Ch. ITP
3. Malignancy	Leukemia Lymphoma	Leukemia Lymphoma
4. Metabolic	-	Gaucher Nieman pick disease
5. Misc	SLE, JIA	Cirrhosis with hypersplenism

APLASTIC /BRUISES

1) ITP
2) Von -willebrand disease
3) Aplastic anemia/Fanconi anemia
4) Pre-leukemic leukemia

HEMOPHILIA

For short case	For Long case
Hemophilia Septic arthritis Trauma Tuberculosis of joint JIA	Hemophilia Severe ITP Platelet function disorders Type 3 vWD Vit K deficiency

ABDOMINAL MASS

Age < 2 yr	Age > 2 yr
Neuroblastoma NHL Rhabdomyosarcoma	Wilm's Tumor Germ cell tumor Neuroblastoma

ENDOCRINE

AMBIGUOUS GENITALIA

Virilised female	Undervirilised males
1. CAH 2. Androgen exposure in utero 3.Maternal tumors	1. Partial androgen insensitivity 2. 5-alpha reductase deficiency 3. CAH 4. Anatomic (gonadal dysgenesis)

OBESITY

obesity with short stature	obesity with N/tall stature
Cushing's syndrome Hypothyroidism GH deficiency Prader-Willi syndrome	Simple obesity Hypothyroidism (on treatment) GH deficiency (on treatment) Klienfelter syndrome

GOITRE

Euthyroid	Thyrotoxicosis	Hypothyroidism
Endemic goiter Simple colloid goiter Euthyroid Hashimoto Subacute thyroiditis Graves' disease on R_x	Graves' disease Hashitoxicosis Subacute thyroiditis	Congenital Hypothyroid Hashimoto Iodine deficiency

PRECOCIOUS PUBERTY

Central (true) PP	Peripheral PP
Idiopathic Intracranial tumours Other CNS lesions Secondary (Steroids, CAH)	Gonadal: (McCune–Albright Tumours) Adrenal: CAH HCG secreting tumours

SHORT STATURE

Proportionate	Disproportionate
Constitutional Familial Malnutrition Chronic illnesses Syndromes Psycosocial deprivation Endocrinopathies Osteogenesis imperfecta	Skeletal dysplasia except OI Congenital Hypothyroidism Rickets

TALL STATURE

Marfan syndrome Homocystinuria Hyperthyroidism Sotos Beckwith–Wiedemann	Klinefelter Kallman CAH Acromegaly

ABDOMINAL EXAM

CLD (Elder child)	CLD (Infant)
1) Wilson's disease 2) Autoimmune hepatitis 3) Drug induced hepatitis 4) Viral hepatitis (elder children only)	1) Biliary atresia 2) Choledochal cyst 3) Neonatal hepatitis 4) PFIC 5) Glycogen storage disorder 6) TORCH infection 7) Tyrosinemia

malformations Hypothyroidism TORCH infections Down/Prader willi IEM Storage disorders	myopathies NMJ disorders (Myasthenia gravis, Botulism)	Meningitis TBM TORCH infections Choroid plexus papilloma	stenosis Dandy Walker Arnold chiari malformation Holoprosencephaly Porencephalic cyst Posterior fossa tumors

MYSTHENIA GRAVIS

1) Autoimmune myasthenia gravis
2) Congenital myasthenic syndromes
3) Toxin-induced myasthenia (Botulism)
4) Organophosphate poisoning

MYOTONIA

1) Myotonic muscular dystrophy
2) Congenital myopathies
3) Atonic CP
4) Hypothyroidism
5) GBS

NEUROFIBROMATOSIS

1) NF-1
2) NF-2
3) Legius syndrome
4) LEOPARD syndrome

SPASTIC PARAPLEGIA

1) Pott's disease/TB spine
2) SOL spine (tumor, abscess)
3) Transverse myelitis

STURGE WEBER SYNDROME

1) SWS
2) Klippel-Trénaunay-Weber's syndrome
3) Beckwith-Wiedemann syndrome
4) Dyke-Davidoff-Masson syndrome

TUBEROUS SCLEROSIS

1) Hypomelanosis of Ito
2) Sturge-Weber syndrome
3) Epidermal nevus syndromes
4) Multiple endocrine neoplasia
5) Isolated brain tumors
6) Cardiac myxoma

CVS

AORTIC REGURGITATION

Aortic Regurgitation (RHD)
Pulmonary Regurgitation

AORTIC STENOSIS

Aortic Stenosis (RHD)
Co-arctation of Aorta
HOCM

ATRIAL SEPTAL DEFECT

ASD
Pulmonary Stenosis

MITRAL REGURGITATION

Mitral Regurgitation (RHD)
Congenital MR
VSD to be ruled out

MITRAL STENOSIS

MS
TS

PATENT DUCTUS ARTERIOSUS

PDA
VSD (subaortic)
AP Window defect
Venous hum

TETRALOGY OF FALLOT

TETRALOGY OF FALLOT

VENTRICULAR SEPTAL DEFECT

VSD

LAST MINUTE REVIEW OF DIFFERENTIALS

CNS

ACUTE FLACCID PARALYSIS

Symmetric	Asymmetric
1. Peripheral neuropathy (GBS) 2.Myelopathy (Transverse myelitis)	1. Polio 2. Traumatic neuritis

CEREBELLAR ATAXIA

Acute	Chronic
Post infectious (Varicella, Coxsackie, Echovirus) Drug induced (Phenytoin, Thallium, Alcohol) Traumatic	Cerebellar Abscess Tumor (Neuroblastoma) DBD Abetalipoproteinemia Congenital Anomalies of Posterior Fossa (Dandy walker, Arnold Chiari, Agenesis of cerebellar Vermis)

CHOREA

1) Sydenham chorea (Rheumatic fever)
2) Wilson's disease
3) SLE
4) Drug induced
5) Cerebral palsy

CEREBRAL PALSY

1) Cerebral palsy
2) Post Meningitic/Kernicteric Sequele
3) Degenerative brain disease
4) Congenital Brain malformation

CVA/STROKE

*Mnemonic: Stroke Says **STITCH ME***

1) **S**ytemic: Sepsis/Dehydration/DKA, Autoimmune / SLE, HUS
2) **T**rauma/ **T**umor : Trauma, Leukemia, lymphoma, SOL
3) **I**nfections : Encephalitis / Meningitis, TBM, Brain Abscess
4) **C**ardiovascular: Cyanotic CHD, MS, AS, IE, Moya Moya, AVM, Vasculitis
5) **H**ematological: ITP, IDA, SCA, Protein C & S deficiency
6) **Me**tabolic: Homocystinuria, Dyslipidemias

DYSTONIA

1) Wilson's disease
2) Drugs
3) Cerebral palsy (Post Kernicteric/Post Asphyxial)
4) SSPE
5) Post Hemiplegic
6) Primary inherited dystonia (e.g. Segawa disease)

LMN FACIAL WEAKNESS

Unilateral	Bilateral
Bell's palsy Trauma Infection (Ramsay Hunt syndrome) Congenital lesion	Myasthenia gravis GBS (Miller Fisher) Myotonic dystrophy

UMN FACIAL WEAKNESS

Unilateral	Bilateral
Stroke Trauma	Cerebral palsy

FLOPPY INFANT

Floppy strong	Floppy weak
Cerebral palsy Cerebral	SMA type 1 Congenital

HYDROCEPHALUS

Communicating	Non communicating/Obstructive
Pyogenic	Congenital Aqueductal

TIPS AND TRICKS FOR SHORT CASES

1) Key to success is: **PRACTICE, PRACTICE, PRACTICE** your scheme
2) Must listen to command & Age of child (If in doubt say, '*Pardon me Sir! Can you repeat the command?*')
3) Seconds count in short case: So be targeted & systematic.
4) Introduction should be brief. *'Asalam o Alaikum (to attendant) ! Mera naam Dr _____ hai aur main apkay bachay ka mooainaa karoon ga/gi.'*
5) Rapport building with child:'*Asalam o alaikum' Beta aap ka kia naam hai?(*Note Hoarseness) *Yeah gift aap ki liyay hai!* (Give gift depending upon age)
6) Bunties, Chocolates, Biscuits etc should not be given as gifts.
7) Keep interacting with child during exam.
8) Be opportunistic: If child is irritable/you think he may cry, auscultate him first and then follow the same routine which you have practiced.
9) Never inflict Pain to Child: A bright student can fail on hurting a child.
10) Describe your findings as you have proceeded. (Be brief as time is short)
11) As the time for this section is short the answers given by the candidate should be precise and relevant to the patient under discussion.
12) Common diseases are common: Make simple differentials, never mention anything rare.
13) Maximum three relevant differentials are enough.(Make Groups if list is long)
14) If examiner asks to tell more differentials then probably you are missing something. RETHINK
15) Always give differentials based upon the findings you have found (even if you have seen the patient)
16) First use generalized description, then specific e.g. cyanotic congenital heart disease likely TOF
17) While presenting short case, skip contradictory findings.
18) Do not use abbreviations
19) Keep hands off the child as long as possible. Focus on Inspection.
20) Narrate while examining the child (Looks professional and impressive)
21) Never show hesitation or pause in examination because examiners will think you are not fluent.
22) Don't use word dysmorphic (offensive) instead describe what you see.

AJ'S HANDY MNEMONICS

INTIAL CHECK LIST BEFORE EVERY SHORT CASE (WIPER)

Wash your hands with sterilizing solution/**W**arm your hands
Introduction & Rapport building
Position the child for exam
Exposure
Right side to approach for examination

FOR SUPPORTIVE TREATMENT OF ANY DISEASE (ALP – OF - TTNM Care)

Admit in Intensive Care Unit
List as per hospital policy + Inform authorities (e.g. WHO team in case of AFP)
Position/ **P**lacement
Oxygen
Fluids
Temp. & others: Tepid sponging + Paracetamol
Tubes: NG, Foley's Catheter
Nutrition: NPO/ supervised/NG Feed
Monitoring (e.g. Vitals, saturation)
Care of comatosed (**EMA's BB Posture**) : **E**ye, **M**outh, Airway, Skin, **B**owel, **B**ladder, **P**osture

LAYOUT OF MANAGEMENT OF ANY CASE

1) MDT (Multi Disciplinary Team Approach)
2) Parental Counselling
3) Treatment of Acute problems
4) Specific treatment
5) Follow up

'PREPARE YOUR WEAPONS'

(PERSONAL EQUIPMENT BOX)

Stationary & Misc	Spiral Pad	2 x Ball points*	Bag/ briefcase	Bait ul Mal forms
	Stop watch (To mange time in long case)			5 x Gift packs (Colors/Cars/ Dolls etc)
Distractions	Toys	Colored ball	Stars	
	Colored Blocks	Crayon	Rattle	
To check Vitals	Stethoscope	Wrist Watch with seconds hand		
	B.P. charts	2 x Inch-tape (steel clamp removed) *		
	2 x B.P. Sets with infant & child cuffs		Growth Charts	2 x Thermometer *
For specific Examination	Otoscope	Ophthalmo-scope	Hammer *	2 x Torches *
	Extra batteries	Cotton wool/wick *	Blunt pins or tooth-picks	Tongue Depressor
	Hand washing solution/Sterilizer		Tissues *	Car key *
	Red topped hat pin for visual field testing in older child Red woolen ball on a thread to test visual field in infants			
	Tuning Fork	Droppers with sweet/saltish fluids/salt & sugar		
	Pair of gloves (Surgical + Polythene)		Soap for smell	

A clean and normally fitting **white Lab coat** is essential component.

Arrange commonly used stuff (indicated by *) in pockets of lab coat.

KNOW YOUR EXAM ASSESSMENT CRITERIA

Note: Basic outline of assessment in FCPS-II examination is used here. Others can follow guidelines as per their specific exam.

SHORT CASES (Total 4 stations) (Time: 10 minutes/station)

Clinical examination skills (5 minutes) 1) Manners 2) Proper & relevant exam (as per instructions) 3) Age appropriate & systematical clinical methods	30 marks
Discussion (5 minutes) 1) Correct findings with logical interpretation & conclusion (25 marks) 2) Justifying diagnosis (20 marks) 3) Appropriate & relevant investigations, management, recent advances (25 marks)	70 marks
Total	100
Passing criteria	60%

LONG CASE (1 station) (Total Time: 70 minutes)

Clinical examination skills (40 minutes) 1) Introduction, informed consent, relevant history 2) Age appropriate & systematical clinical methods to elicit clinical findings (Detailed examination of relevant system)	20 marks
Case presentation (15 minutes) 1) Presentation skills (5 marks) 2) Correct findings (10 marks) 3) Logical interpretation of findings (15 marks) 4) Differential diagnosis (10 marks)	40 marks
Discussion (15 minutes) 1) Justified & relevant investigations (10 marks) 2) Management plan + Rehabilitation (15 marks) 3) Prognosis & counseling (10 marks) 4) Relevant recent advances/ complications (5 marks)	40 marks
Total	100
Passing criteria	60%

CONTENTS